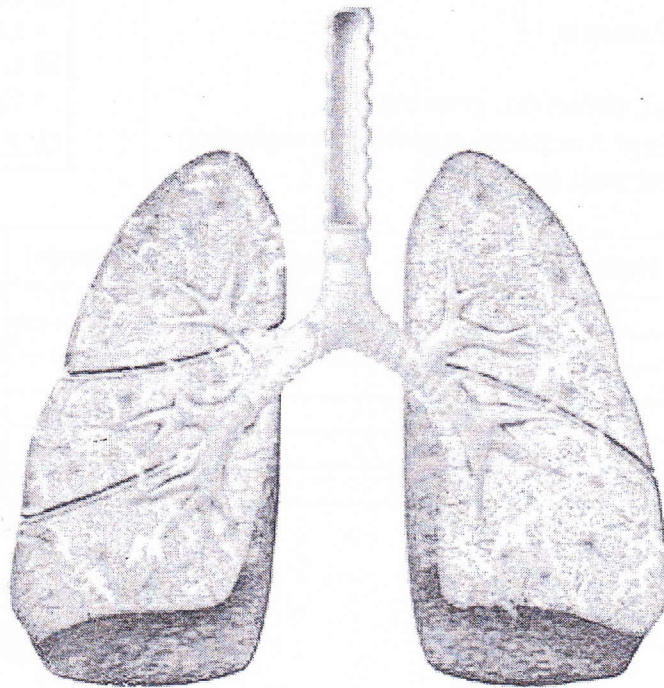




Pediatric Pulmonology

By

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Chest Examination

A) Inspection

1. Shape: Pigeon, Barrel-shaped, Pectus excavatum
2. Symmetry:
3. Respiratory movements
 - Rate
 - Working ala nasi, retraction, grunting
 - Work of breathing: Accessory muscles of respiration
 - Paradoxical chest wall movements

Symmetry:

- ☒ Unilateral bulge
 - Effusion, Pneumothorax
- ☒ Unilateral retraction
 - Collapse, Fibrosis

How to DD? Resp. movements

	Heart Rate (min)	Respiratory Rate (min)	Blood Pressure
Newborn	100-150	60	80 / 50
6 months	120	50	80 / 60
1-5 year	120	40	90 / 60
5-8 years	100	30	100 / 70
10 years	90	20	100 / 70
≥ 12 years	90	12-18	110 / 70

B) Palpation

1. Mediastinal position
 - Heart
 - Trachea
2. Chest expansion
3. TVF

TVF/VR	Pathology
↑↑	Consolidation
↓↓	Effusion/Collapse

C) Percussion

- Limited value in small infants

Note	Pathology
Resonant	Normal
Hyperresonant	Emphysema, Pneumothorax
Dull	Consolidation, Collapse
Stony dull	Effusion

D) Auscultation

- Breath sounds: Vesicular, Harsh vesicular, Bronchial
- Adventitious sounds: Wheezes, Crepitations, Rub
- Conducted upper airway sounds are the most common cause of false +ve chest findings
- D'Espine sign: Bronchial breathing below T4 vertebra (T2 spine) Cause: Mediastinal LN

Non-pulmonary causes of clubbing:

- Cardiac: Cyanotic CHD, IE
- Pulmonary: Chronic hypoxia
- Hematologic: Thalassemia
- GIT: IBD (UC, CD), small bowel lymphoma, polyposis coli, liver cirrhosis
- Unilateral clubbing:
 - Vascular disorders (Subclavian arterial aneurysm, brachial AVF)
 - Median nerve injury
 - Shoulder subluxation
 - Local trauma

Investigations of Chest Diseases

A) Laboratory

- ☒ **Blood**
 - **CBC:** Hb (anemia of chronic disease), WBC (↑↑ in infection, ↓↓ in immunodeficiency) & PLT (Collagen-vascular diseases)
 - **ESR & CRP:** ↑↑ Chest infection (TB)
 - **Arterial blood gases (ABG)**
 - **ANCA** (Anti-neutrophil cytoplasmic Ab): Wegener granulomatosis & Churge-Strauss
 - **Anti-centromere Ab:** in scleroderma
- ☒ **Sweat chloride test:** Cystic fibrosis
- ☒ **Microbiology**
 - **Sample:** Sputum, ETT, stomach wash, fasting gastric aspirate, BAL or lung biopsy
 - **Examination**
 - Cytological: ↑↑ PNLs (Bacterial), ↑↑ Lymphocytes (TB & viral), Malignant cells
Hemosiderin granules in macrophages (Hemosiderosis)
Lipid laden macrophages in aspiration pneumonia
Direct smear for bacteria & fungi
 - Culture
- ☒ **Urine**
 - **Urinalysis & Urinary protein/creatinine ratio** ($N < 0.2$): Proteinuria, when?
 - **Urine culture & sensitivity**

B) Imaging

- ☒ **CXR (PA & Lateral views):**
 - **Lung:** Pulmonary congestion & infection
 - **Heart:** Cardiomegaly
- ☒ **Upper airway films**
 - **Steeple sign:** Subglottic tracheal narrowing (Inverted V appearance)
 - **Thumb sign:** Epiglottitis
- ☒ **CT & MRI:** Mediastinal lesions, bronchiectasis & cystic parenchymal diseases
- ☒ **Fluoroscopy**
- ☒ **Sinus:** X-rays & CT
- ☒ **Chest transillumination:** Infants < 6 months (*pneumothorax*)
- ☒ **Barium swallow**
- ☒ **Bronchography:** Bronchiectasis
- ☒ **Angiography;** Vascular ring
- ☒ **Radionucleotide Lung scan (Isotopic scan)**
 - Assessment of ventilation & perfusion
 - Diagnosis of pulmonary embolism & congenital vascular anomalies

C) Pulmonary function tests

- ☒ **Spirometry:** measurement of VC and its subdivisions and expiratory flow rates
- ☒ **Plethysmography:** measurement of TLC, Airway resistance
- ☒ **Diffusing capacity for carbon monoxide (DLCO):** measurement of diffusion
- ☒ **Exercise testing**

D) Polysomnography

- It is the gold standard test for obstructive sleep apnea or hypoventilation
- pH probe studies are can be added (GERD)

E) Invasive:

☒ Endoscopy

- Laryngoscopy: Direct & indirect
- Bronchoscopy:
 - Diagnostic: Recurrent pneumonia, suspected FB, hemoptysis
 - Therapeutic: Removal of FB & Bronchoalveolar lavage (BAL)

☒ Thoracoscopy

☒ Lung biopsy: Open or transbronchial

☒ LN biopsy

☒ Thoacocentesis: Obtaining fluid from the pleural cavity

Needle puncture should be along the upper border of the rib, why?

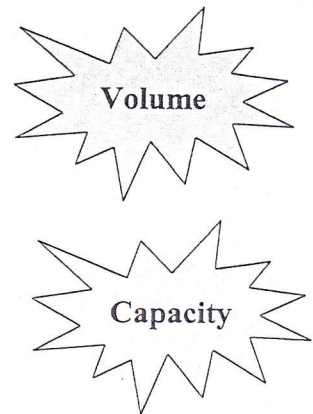
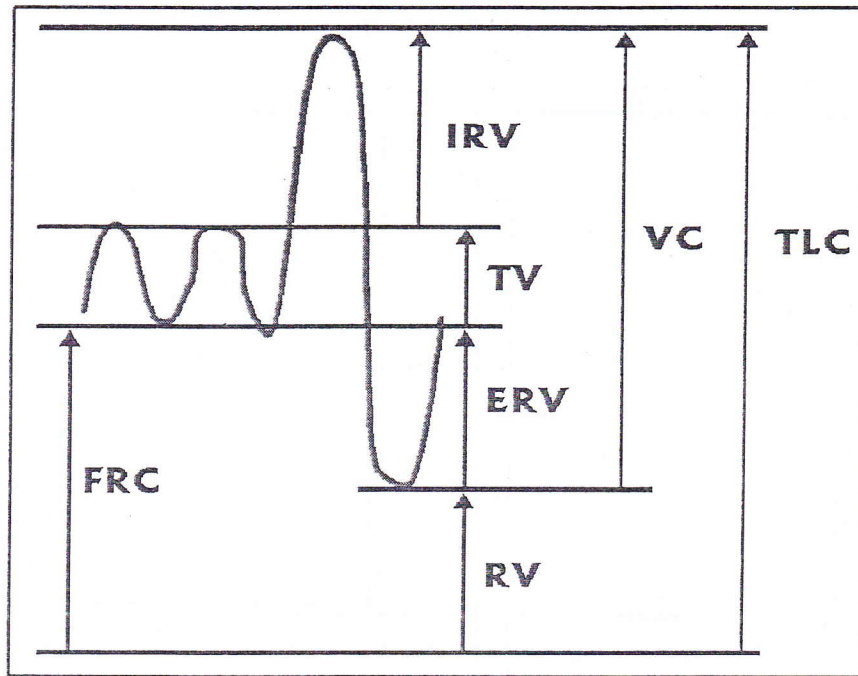
Complications: Infection, bleeding, pneumothorax, liver/spleen injury

Pleural fluid examination (Physical, Chemical, Cytological & Bacteriological)

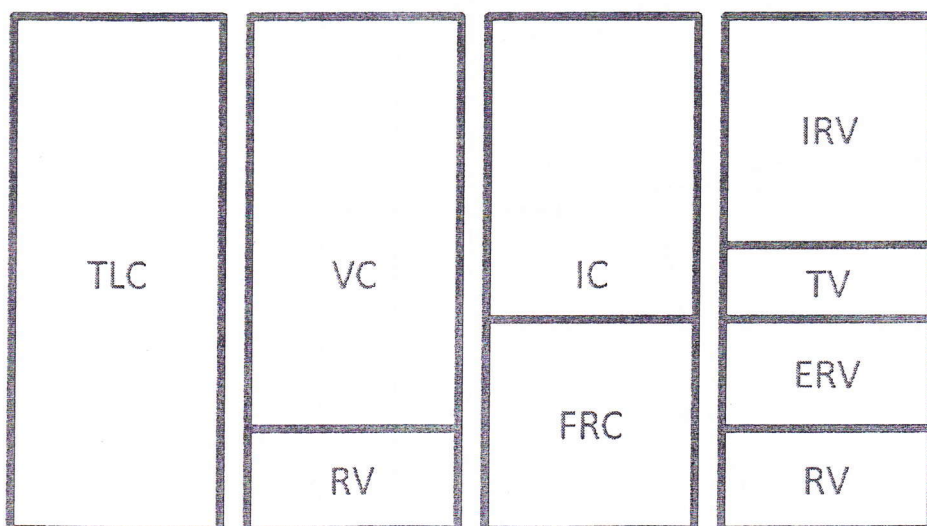
- Physical: Clear in transudate, turbid in others
- Cytological: ↑↑ PNLs (Exudate), ↑↑ Lymphocytes (TB), Malignant cells
- Chemical: Milky fluid with ↑↑ TAG (Chylous), blood-stained (Tumors)
- Bacteriological: Culture

	Transudate	Exudate
Aspect	Clear	Turbid
Specific gravity	Low (< 1015)	High (> 1015)
Proteins	Low (< 2.5 g/dl)	High (> 2.5 g/dl)
Cell Number	Few (< 2000 /mm ³)	Many (> 2000 /mm ³)
Cell Type	Lymphocytes	PNLs
LDH	Low (< 200 IU/L)	High (> 200 IU/L)

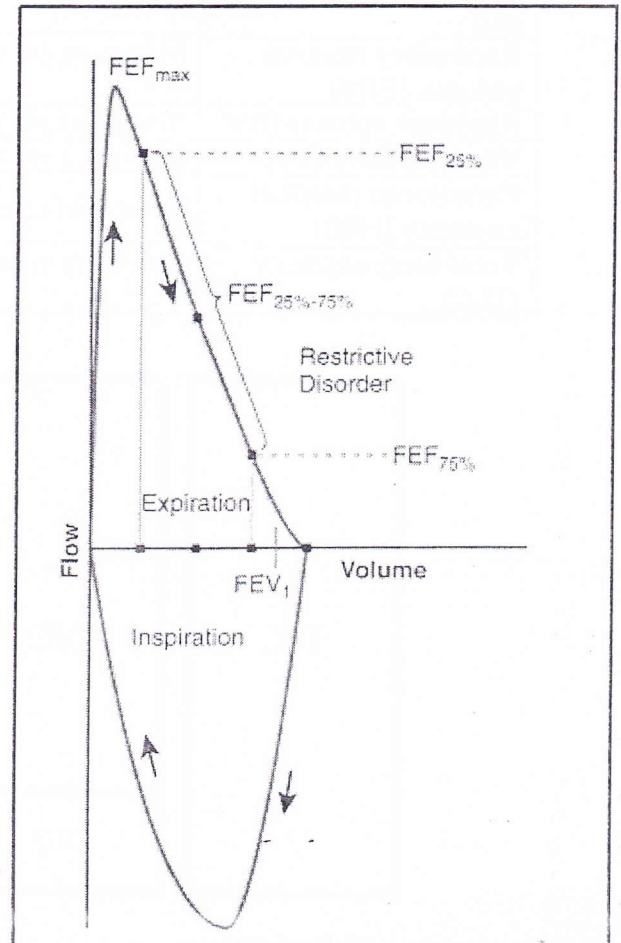
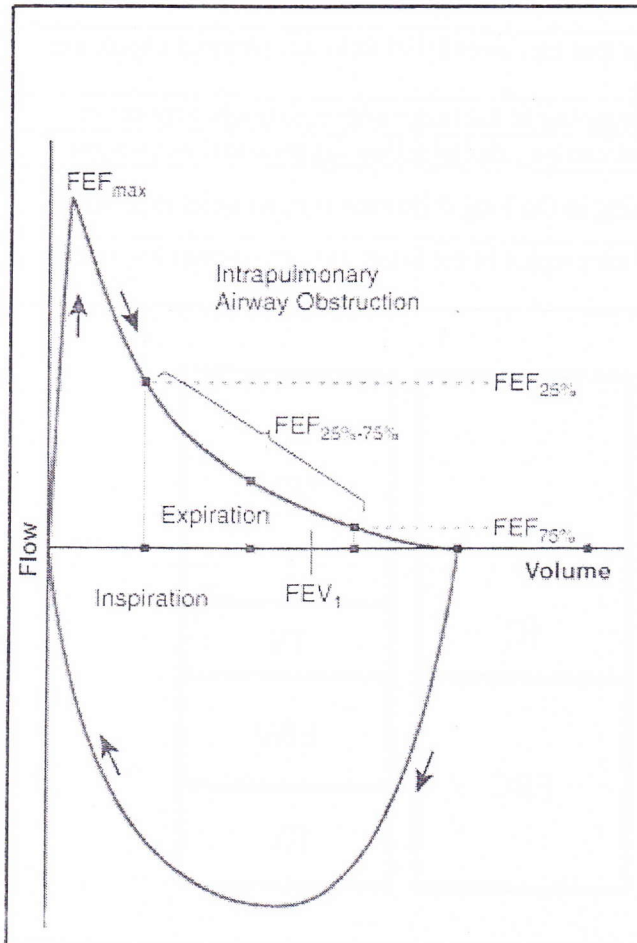
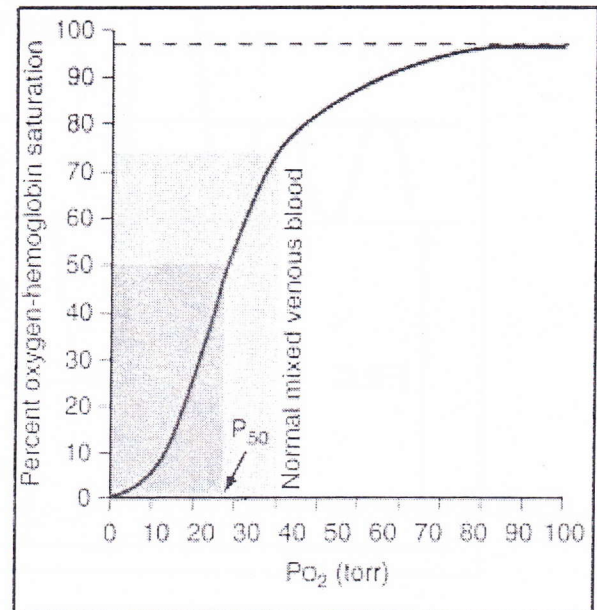
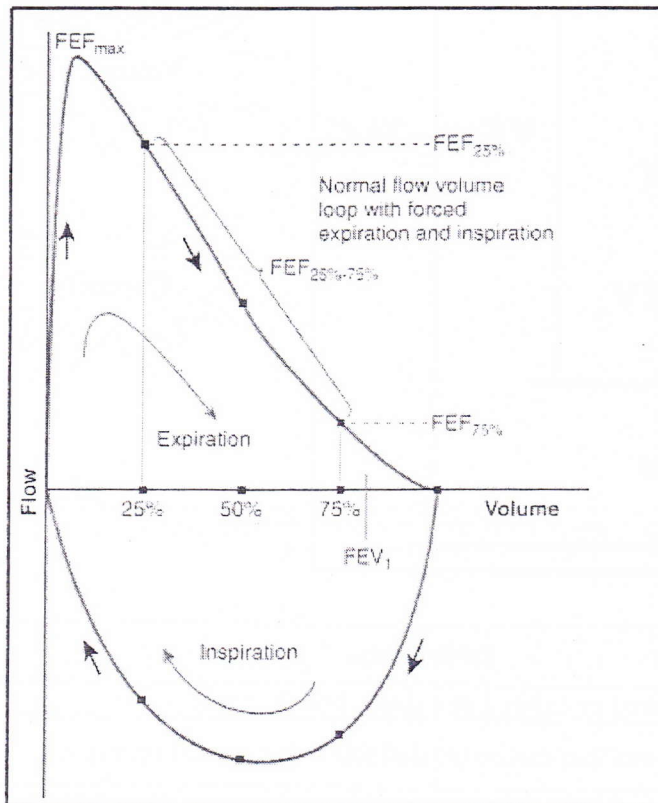
Lung Volumes & Capacities



Volume/ Capacity	Definition
Tidal volume	Volume of air inspired or expired in a single breath at rest
Inspiratory reserve volume (IRV)	Maximum gas volume that can be inhaled following normal inspiration
Inspiratory capacity (IC)	Amount of air inspired by maximum inspiration after normal expiration
Expiratory reserve volume (ERV)	Maximum gas volume that can be exhaled following normal expiration
Residual volume (RV)	The volume of gas remaining in the lungs after maximum expiration
Vital capacity (VC)	Maximum volume that can be exhaled following maximal inspiration
Functional residual capacity (FRC)	Volume of air remaining in the lung following normal quiet expiration
Total lung capacity (TLC)	Maximum volume of air present in the lungs after maximum inspiration



Flow Volume Loops



Causes of Recurrent or Persistent; Cough in Children

1. Asthma

- Aspiration
- Alpha-1-Antitrypsin deficiency
- Aspergillosis

2. Bronchitis

- Bronchiolitis
- Bronchiolitis obliterans
- Bronchopneumonia
- Bronchopulmonary dysplasia
- Bronchiectasis
- Pneumonitis
- Pertussis
- Poisoning (Organophosphorous)
- Post-nasal discharge (Sinusitis)

3. Cystic fibrosis

- Ciliary dyskinesia
- Cardiac (Heart failure)
- Cricoid in coordination
- Cacinoid syndrome
- Chronic renal failure
- Compression 

Compression:

- **Lumen:** FB, secretion, tumors
- **Wall:** Tracheomalacia, bronchomalacia, tumors
- **Outside:** LN, cysts, vascular ring, tumors

4. Endobronchial (TB & tumors)

- Esophageal atresia (TOF)

5. Foreign body inhalation

- Fungal infection

6. GERD

7. Heart failure

- Hiatus hernia
- Hemosiderosis
- H-shaped tracheo-esophageal fistula
- Habit cough (Psychogenic cough)

8. Immunodeficiency

- Immotile cilia syndrome
- Irritation: Smoking
- Irritation of the external auditory canal

Causes of Cough

A) Acute cough (Duration < 2 wks)

- a. **With RD:** Acute bronchiolitis, Acute asthmatic attack, Pneumonia
- b. **Without RD:** Acute bronchitis, Acute laryngitis, Acute sinusitis

B) Prolonged cough (2 wks - 2 months)

- a. Complicated bronchitis (Bacterial bronchitis, pneumonia, segmental collapse)
- b. Acute sinusitis
- c. Pertussis & pertussis-like illness

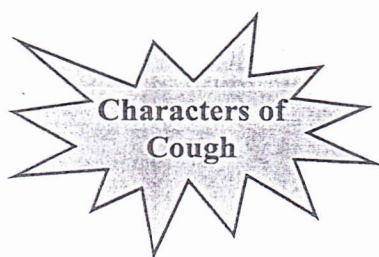
C) Chronic cough (Duration > 2 months)

- Chronic infections: TB, bronchiectasis
- Immunodeficiency
- FB inhalation
- Persistent asthma
- Recurrent aspiration: GERD, TOF
- Congenital abnormality of lung and heart
- Cystic fibrosis
- Bronchopulmonary dysplasia

Diagnostic Approach for Cough

A) History

- ☒ Character of cough
- ☒ History suggestive of FB inhalation: **Sudden** onset of cough, chocking & cyanosis
- ☒ Recurrence & seasonal variation: Asthma
- ☒ TB toxemia: High fever, night sweating, loss of weight, loss of appetite
- ☒ Aspiration
- ☒ Smoking & passive smoking
- ☒ Nasal obstruction & nasal discharge
- ☒ Other systems: GIT symptoms (CF), Cardiac (HF)
- ☒ Animal exposure: Chlamydia (birds), Yersinia (rodents), Q fever (cattle & sheep)



Character	Diagnosis
Productive	Bronchitis, bronchiectasis
Brassy (Metallic)	Tracheitis
Bovine (Barking)	Acute laryngitis
Tight (Wheezy)	Asthma (Reactive airways)
Nocturnal	Asthma, sinusitis
Early morning	Asthma & bronchiectasis
With vigorous exercise	Asthma (exercise-induced)
Disappears with sleep	Habit cough
Paroxysmal	Pertussis
Staccato	Chlamydia pneumonia

B) Examination

a. General

- Pallor: anemia (anemia of chronic disease or hemosiderosis)
- Failure to thrive, clubbing & cyanosis

b. Chest examination

- Inspection, palpation, percussion & auscultation

C) Investigations: See before

Bronchial Asthma

Definition

- Chronic diffuse **inflammatory** obstructive disorder of the airways characterized by
 - Airway hyper responsiveness (AHR)
 - High degree of reversibility either spontaneously or with treatment
- Bronchial asthma is characterized by recurrent episodes of wheezing, breathlessness, chest tightness, and coughing, particularly at night or in the early morning
- The majority of asthmatic children have allergic component
- Asthma can be effectively controlled, although it cannot be **cured**

Epidemiology

Prevalence of BA is ↑↑

- 10% of school children
- 80-90% of asthmatics are symptomatic before the age of 6 yrs
- Of all young children with recurrent wheezes, only a minority will have persistent asthma
- Children living in rural areas & farming communities are less likely to develop asthma and allergy (**Hygiene theory**)
- **Risk** for development of **persistent asthma**:
 - Parental asthma
 - Tobacco exposure
 - Wheezes without cold
 - Allergy (atopic dermatitis, allergic rhinitis, food allergy)
 - Male sex
 - Low birth weight
 - Reduced lung function at birth
 - Chlorinated swimming pools
 - Use of paracetamol

Etiology (Multifactorial)

- Environmental exposure (Asthma triggers)
- Genetic: Multiple genes causing asthma & asthma-related traits (atopy, atopic dermatitis...)

Patterns of recurrent wheezing in childhood (Clinical pattern)

A) Transient early wheezing

- Recurrent wheezes triggered by viral infection
- Preschool years & resolves by 5 years of age
- No asthma later in life

B) Persistent atopy-associated asthma

- IgE-mediated
- Associated with atopy; atopic dermatitis, allergic rhinitis, food allergy
- Highest risk for persistent asthma

C) Non-atopic wheezing

- Recurrent wheezes triggered by RSV
- Resolves in late childhood
- No increased risk for persistent asthma

D) Late-onset asthma in females associated with obesity & early-onset puberty

- Onset: 8-13 yrs

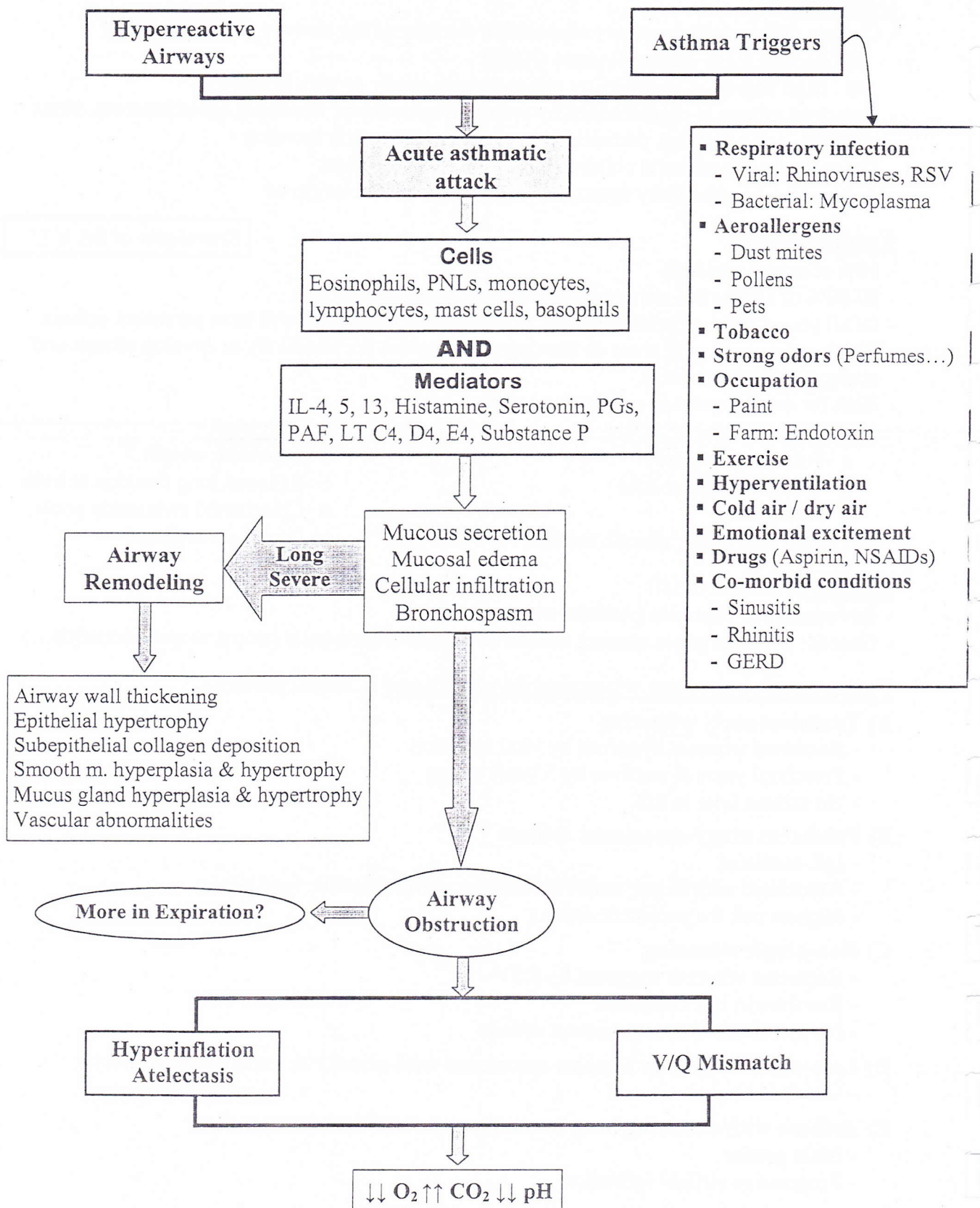
E) Asthma with declining lung function

- Male gender
- Progressive airflow limitation

F) Occupational-type asthma

- Farms: Endotoxin exposure

Pathogenesis



Clinical Picture (It is a clinical diagnosis)

Markers of atopy

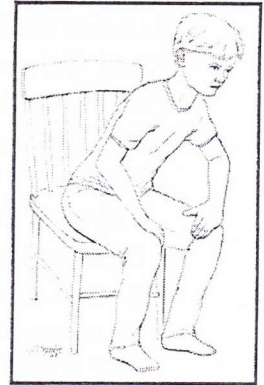
A) Symptoms

- Recurrent, intermittent dry **cough** (Nocturnal &/or early morning)
- Cough or wheezes after exercise
- Cough or wheezes after exposure to airborne allergens
- **Atopic March:** Infants with atopic dermatitis → Allergic rhinitis → Asthma
- Co-morbid conditions: can mimic or worsen asthma

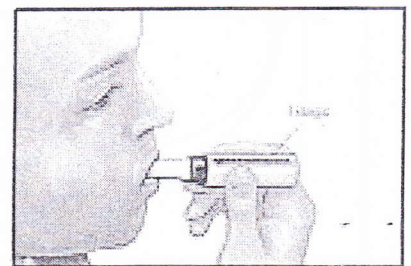
Atopic March

B) Signs

- Vesicular breath sounds with prolonged expiration
- Expiratory **wheezes** (Usually sibilant)
- Variable degrees of **RD** (Tripod position) ⇒
- Chest Ex.: with prolonged expiration, expiratory wheezes
- **Severity** of asthma attacks: Mild, Moderate or Severe



	Mild	Moderate	Severe	Imminent respiratory arrest
Breathlessness	Walking	Rest	Rest	
Talking	Sentences	Phrases	Words	
Alertness	Agitated	Agitated	Agitated	Drowsy
RR	↑↑	↑↑	↑↑	
Pulse	< 100	100-120	> 120	Bradycardia
Pulsus paradoxus	Absent	May be present	Usually present	Absent
Accessory muscles	No	Yes	Yes	Paradoxical
Wheezes	Yes	Yes	Yes	Absent
PEF	> 70%	40-70%	< 40%	< 25%
PaO ₂	Normal	> 60 mmHg	< 60 mmHg	
PaCO ₂	< 42 mmHg	< 42 mmHg	> 42 mmHg	
SaO ₂	> 95%	90-95%	< 90%	

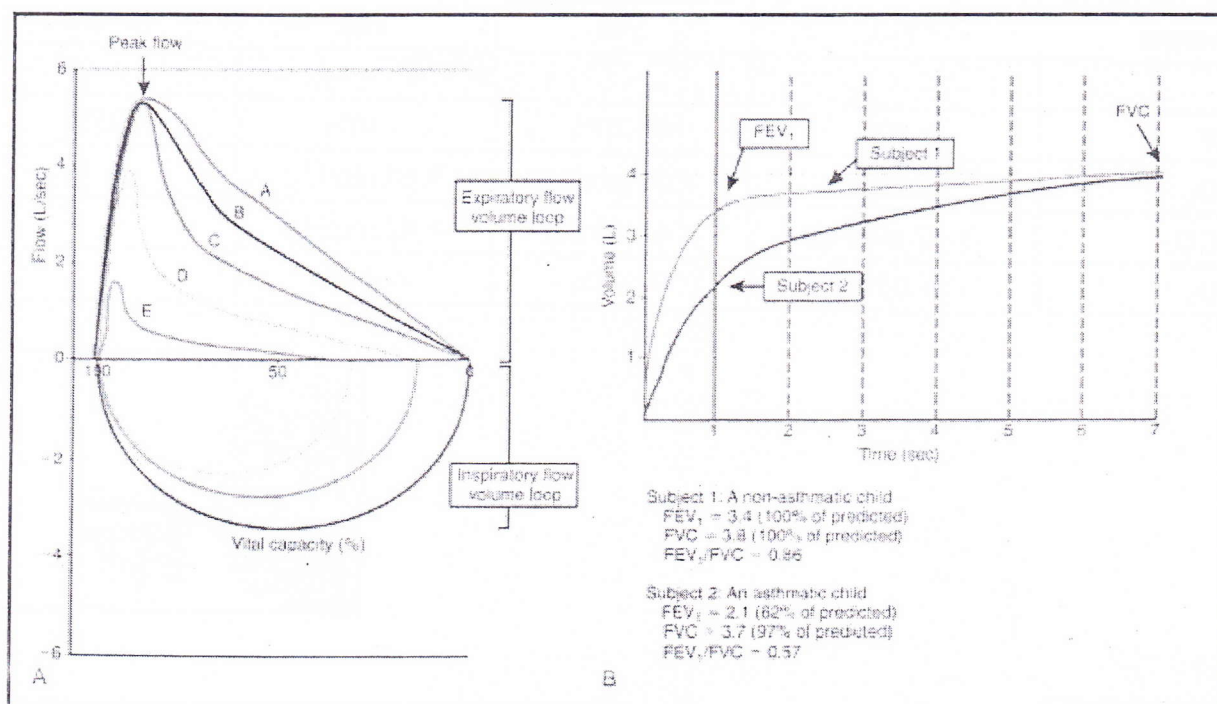


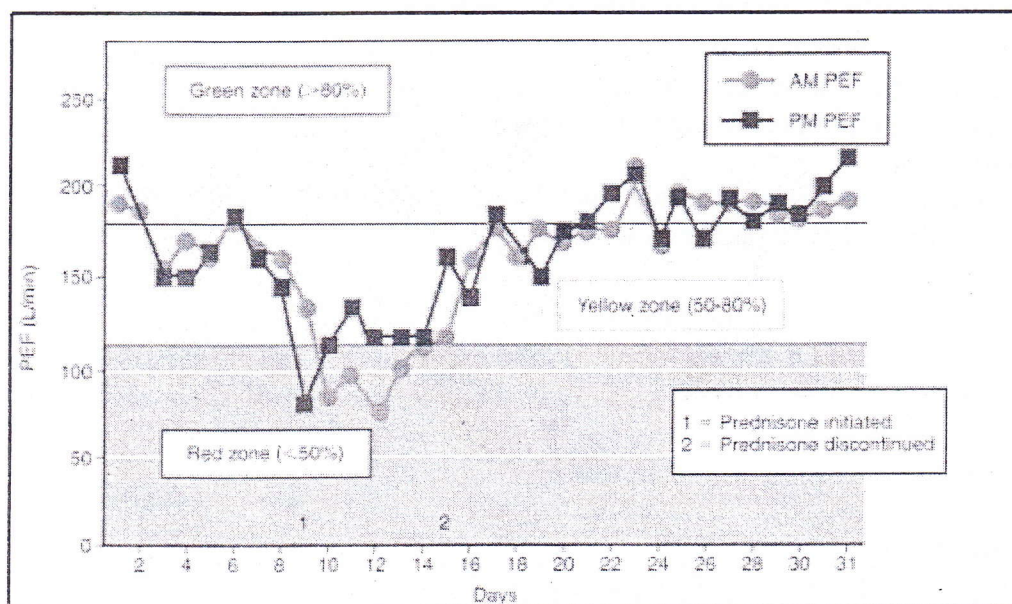
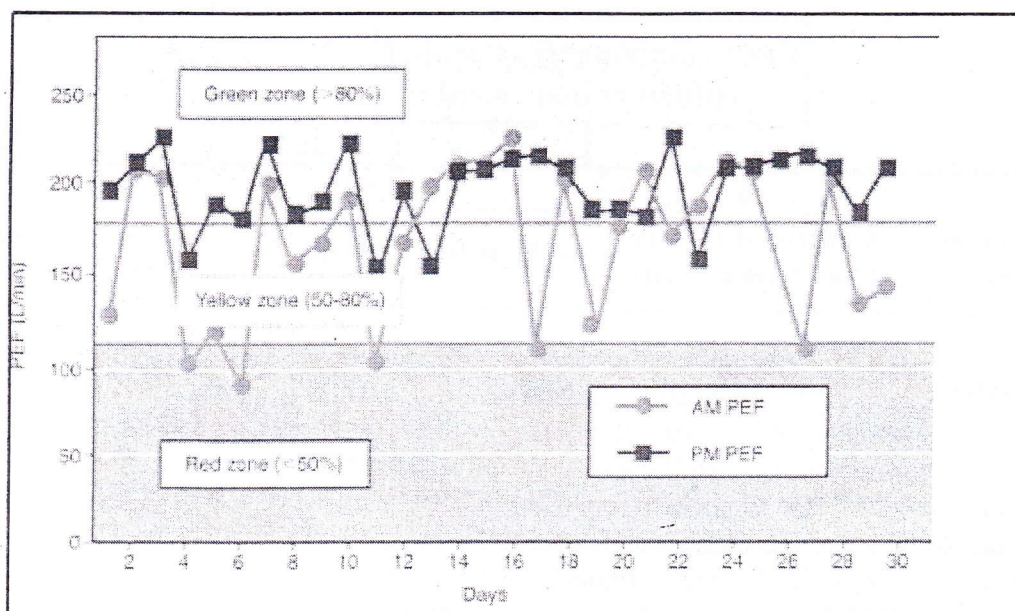
Investigations

- CBC ($\uparrow\uparrow$ Eosinophils)
- $\uparrow\uparrow$ Serum IgE (Total & specific)
- ABG
- Prick skin test
- Inhalation bronchial challenge (*Methacholine or histamine*)
- Exercise challenge (*Running for 6-8 minutes*)
- FENO: Exhaled NO "Marker of airway inflammation"
- CXR: Hyperinflation
- Pulmonary function tests (**Spirometry**): *Feasible in children > 6 yrs*
 - FEV₁: $\downarrow\downarrow$
 - FEV₁/FVC: $\downarrow\downarrow < 80\%$
 - Bronchodilator response to inhaled β_2 agonists: $\uparrow\uparrow$ FEV₁ $\geq 12\%$
 - Exercise challenge: $\downarrow\downarrow$ FEV₁ $\geq 15\%$
 - Variation in FEV₁ or PEF $\geq 20\%$
 - PEF (Daily): 3 zones (Green "80-100%", Yellow "50-80%" & Red "<50%")

Parameter	Obstructive	Restrictive
FVC	Normal or Decreased	Decreased
FEV ₁	Decreased	Decreased
FEV ₁ /FVC	Decreased	Normal (or Increased)
FEF ₂₅₋₇₅	Decreased	Normal
TLC	Normal or Increased	Decreased
RV	Increased	Normal or Decreased
RV/TLC	Increased	Normal or Decreased

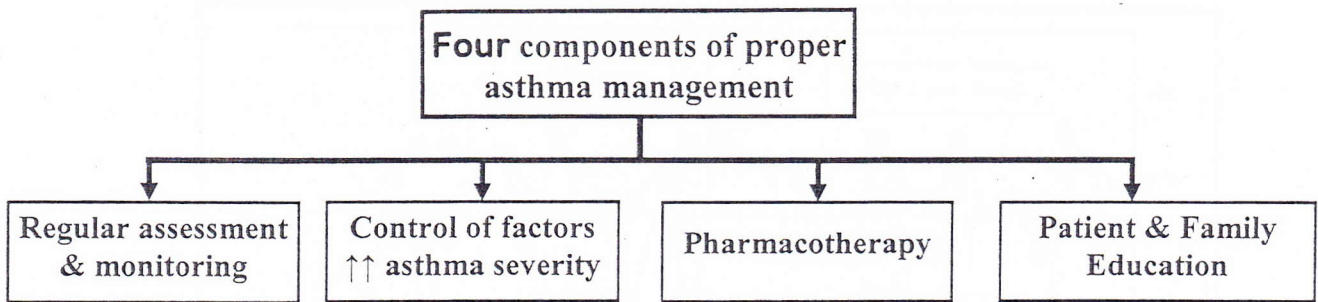
FEV₁: Forced expiratory volume in the 1st sec.
FVC: Forced vital capacity





Treatment of Bronchial Asthma

(National Asthma Education & Prevention Program)



A) Regular assessment & monitoring

- Assessment of asthma severity: Mild or persistent (Mild, moderate, severe)
- Assessment of asthma control: Well-controlled, not well-controlled, very poorly controlled
- Responsiveness to therapy (& medications side effects)
- Check-up: 2-4 / year are recommended (Items to be checked??)
- Spirometry is recommended at least annually
- PEF is recommended daily (preferably in the morning)

B) Control of factors ↑↑ severity of asthma

Egg-allergy!

- ↓↓ Environmental exposure (e.g., annual Influenza vaccine...)
- Rx of co-morbid conditions (GERD, Rhinitis, Sinusitis)

a. GERD

- Occurs in 60% of asthmatic patients
- Mechanism: Aspiration & vagal stimulation
- Investigation:
- TTT: Positioning, dietary management, Drugs (Antacids & H₂ blockers, proton-pump inhibitors) & rarely surgical management

b. Rhinitis

- Occurs in 90% of asthmatic patients
- TTT: Avoidance of offending allergens, oral antihistaminics, nasal steroids

c. Sinusitis

- Occurs in 60% of asthmatic patients
- TTT: Antibiotics, nasal saline irrigation, nasal steroids

C) Pharmacotherapy

- Quick-relief, controller therapy & stepwise approach
- Rx of asthma exacerbation

D) Patient & family education

- Basic facts about asthma
- Medications side effects
- Goals of management
- Environmental asthma triggers
- Written management plan (Daily management & Rx of exacerbations)
- Check adherence to Rx
- Regular F/U visits

Management of Asthma Exacerbations

Quick-relief medications

1. Short-acting inhaled β_2 -agonist (SABA)

☒ Salbutamol

- Inhalation (100 μ g/puff): 2 puffs every 20 min
- Nebulization (5mg/ml): 0.5 ml + 2-3 ml NS every 20 min "0.15 mg/kg"
- IV: 0.2 μ g/kg/min. (max. 4 μ g/kg/min)
- Oral (Ventoline 2mg/5ml): 0.6 mg/Kg/day

☒ Terbutaline, albuterol, levalbuterol

2. Anti-cholinergic agents

☒ Ipratropium

- Inhalation (20 μ g/puff): 2 puffs every 6 hr
- Nebulization (0.5 mg /2 ml): every 6 hr

3. Short course of systemic steroids (Methylprednisolone, prednisone, prednisolone)

Dose: 1-2 mg/Kg/day for 3-7 days

4. Adrenaline (IM or SC): 0.01 mg/Kg/dose (*Can be repeated after 15-30 minutes*)

Side effects of SABA:

1. Tachycardia
2. Tremors
3. Hypokalemia
4. Hypoxemia, why?

Asthma exacerbations:

- Acute episodes of worsening of symptoms
- V/Q mismatch: Hypoxemia
- Time: Usually 12 am- 8 am
- Complications: Air leak (Pneumomediastinum & pneumothorax)
- **Status asthmaticus:** Severe exacerbation **not** responding to standard Rx
- **Risk factors for asthma morbidity & mortality:**

- | | |
|---|------------------------------|
| o Previous asthma exacerbations | o Poverty |
| o ≥ 2 Hospitalization in the past yr | o Crowding |
| o Poor response to systemic steroids | o Mother < 20 yrs |
| o Male sex | o Inadequate medical care |
| o Low birth weight | o No regular F/U |
| o Urban environment | o Lack of action plan |
| o Tobacco exposure | o Delay in care |
| o Antigen exposure | o Alcohol or substance abuse |

A) Home management

- All children with asthma should have a written action plan ($\downarrow\downarrow$ Mortality)
- Initial Rx: Inhaled SABA (3/hr)
 - Good response (= Symptoms subside + PEF > 80%):
SABA every 4 hr + contact physician
 - Incomplete response (= Symptoms improve + PEF 50-80%):
SABA + Oral steroids (prednisone) + Consult physician
 - Poor response (= Symptoms persist + PEF < 50%):
SABA + Oral steroids (prednisone) + Transport to ER
NB: Injectable form of adrenaline (Life-threatening conditions)

B) ER management

- **History taking:** Onset, triggers, medications received, current Rx, previous attacks
- **Assessment:**
 - **Clinical:** Vital signs, mental status, breathlessness, RD(4), accessory muscles, chest expansion, air entry, wheezes, silent chest, pulsus paradoxus
 - **Laboratory:** Oxygenation (pulse oximetry, ABG), PFTs (FEV₁ or PEF)
- **Risk factors for morbidity & mortality:** *See before*
- **Management:**
 - Oxygen therapy: Mask or prongs
 - Inhaled SABA (every 20 min) ± Ipratropium
 - Systemic steroids (Oral or IV)
 - IM adrenaline may be given in severe cases

C) Hospital management

- **Indications:** Failure of response within 1-2 hrs of intensive Rx Or high risk of mortality
- **Assessment:**
 - **Clinical:**
 - **Laboratory:** Oxygenation (pulse oximetry, ABG), electrolytes, CBC, CXR
- **Management:**
 - Oxygen therapy: Mask or prongs
 - Inhaled SABA (may be continuous) ± Ipratropium
 - Systemic steroids (IV or oral)
 - IV Theophylline
 - IV β_2 -agonist
 - IM or SC adrenaline: 0.01 mg/Kg/dose (*Can be repeated after 15-30 minutes*)
 - IV MgSO₄
 - Inhaled heliox
 - Hydration status: Dehydration Vs the risk of SIADH
 - ETT & Mechanical ventilation: may be needed in cases of respiratory failure
 - Mucolytics & chest physiotherapy should be **avoided**, why?

Mechanical ventilation in severe asthma

- Mechanical ventilation should be anticipated
- Elective intubation is safer than emergency intubation
- Careful balance of enough pressure:
 - Low pressure → Hypoventilation (airway obstruction)
 - High pressure → Hyperinflation, Air leak (Barotrauma)
- Settings:
 - ↑↑ Expiratory time (I/E = 1:3 or even 1:4)
 - ↓↓ PEEP ≤ 4 cm H₂O (Zero-PEEP may be used)
- Myopathy in asthma: Hopkins syndrome, steroids, fatigue

Discharge (*Stabilization*)

- Normal physical findings
- Oral intake
- PEF > 70%
- O₂ saturation > 92% (room air) for ≥ 4 hr
- Rx on discharge: Inhaled β_2 -agonist (up to every 3-4 hrs) & steroids (for 3-7 days)
- F/U visits are planed after 1-2 wks
- Stepwise adjustment of controller Rx

Outcome Mortality is rare in medical centers (Most deaths occur at home)

Prevention of asthma

- Reduce exposure to major allergens
- Avoidance of environmental tobacco smoke
- Prolonged breastfeeding
- No "solid" foods for the first 6 months
- Egg and fish only after 12 months of age
- Maternal elimination diet during breast feeding
- Avoid day-care centers during infancy
- Immunizations: All vaccines including varicella & influenza
- Hygiene theory: Microbial exposure in early life may ↓↓ asthma incidence (Rural areas)

Prognosis

- Of all young children with recurrent wheezes, only a minority will have persistent asthma
- In children with mild asthma, the **remission** rate is $\approx 50\%$
- In children with severe asthma, $\approx 95\%$ become asthmatic **adults**
- **Risk** for development of **persistent asthma**

Causes of Wheezes

Definition

Single (Non-recurrent) Wheezes	Chronic (Recurrent) Wheezes
Acute bronchiolitis	Bronchial asthma
Severe bronchopneumonia	Recurrent aspiration
FB inhalation	FB inhalation
Organo-phosphorous poisoning	Chronic infection
HF	HF
Transient bronchial hyperreactivity	Bronchopulmonary dysplasia
Asthma (1 st attack)	Bronchiolitis obliterans
	Airway compression

Assessment of Asthma Severity

- For patients not on controller
- Assign to the most severe
- Initiate higher level controller

Severity	Impairment						Risk
	Daytime symptoms	Nighttime awakening	Interference with activity	SABA use	FEV1 % predicted	FEV1/FVC ratio	Exacerbation requiring Systemic steroids
Mild intermittent	≤ 2/wk	≤ 2/month	No	≤ 2 d/wk	> 80%	> 80%	0-1/ yr
Mild persistent	> 2/ wk	> 2/ month	Minor	> 2 d/ wk	> 80%	> 80%	≥ 2/ yr
Moderate persistent	Daily	> 1/wk	Some	Daily	60-80%	75-80%	≥ 2/ yr
Severe persistent	Throughout the day	Frequent	Extreme	Several/d	< 60%	< 75%	≥ 2/ yr

Recommended Step for initiating Rx: Step 1, 2, 3, 4

Age groups: 0-4 yrs, 5-11 yrs, ≥ 12 yrs

Classification of Asthma Control

- For patients on controller
- Assign to the most severe

Severity	Impairment						Risk	
	Daytime symptoms	Nighttime awakening	Interference with activity	SABA use	FEV1 % predicted	FEV1/FVC ratio	Exacerbation requiring Systemic steroids	Drug SE
Well-controlled	≤ 2/wk	≤ 2/month	No	≤ 2 d/wk	> 80%	> 80%	0-1/ yr	
Not well-controlled	> 2/ wk	> 2/ month	Some	> 2 d/ wk	60-80%	75-80%	≥ 2/ yr	
Very poorly controlled	Throughout the day	> 1/wk	Extreme	Several/d	< 60%	< 75%	≥ 2/ yr	

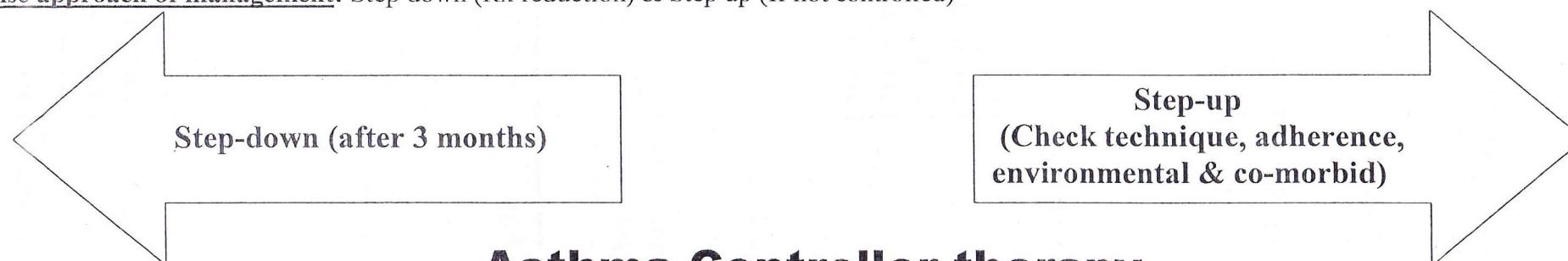
Well-controlled: Keep on, consider step-down if well for ≥ 3 months **Not Well-controlled:** Step-up, assess after 4-6 wks
Very poorly controlled: Short course of systemic steroids, Step up (1-2 steps) & assess in 2wks

Stepwise Approach for Management of Asthma

	Step 1	Step 2	Step 3	Step 4	Step 5	Step 6
Preferred	SABA prn	Low-dose ICS	Medium-dose ICS	Medium-dose ICS & LABA	High-dose ICS & LABA	High-dose ICS & LABA & Oral steroids
Alternative	-	Cromolyn or LTRA	Low-dose ICS & LTRA	Medium-dose ICS & LTRA	High-dose ICS & LTRA	High-dose ICS & LTRA & Oral steroids

Quick Relief (All patients): Short-acting β_2 agonists (inhaled or oral): Salbutamol, Terbutaline $\pm$ Ipratropium ($\pm$ Short courses of systemic steroids)

Stepwise approach of management: Step down (Rx reduction) & Step up (If not controlled)



Asthma Controller therapy

Inhaled Steroids	LABA	Leukotriene Modifiers	Other asthma controllers
1. Beclomethasone (Becotide) 2. Budesonide (Pulmicort) 3. Fluticasone (Flixotide) 4. Triancinolone 5. Fluticasone + Salmeterol (Seretide) Dose: 100 μ g/dose bid or tid Forms: Inhaler or Diskus	Salmeterol (Serevent) Dose: 20 μ g/dose bid	Montelukast (Singulair 5 & 10 mg tab) Dose: 5-10 mg oral single daily dose (when?)	▪ Ketotifen (Zaditen): 0.05 mg/Kg/day bid ▪ Cromolyn Na (Intal inhaler): 5 mg/dose qid ▪ Sustained-release theophylline (Theo SR): 15-20 mg/kg/day bid ▪ Oral steroids (Prednisone): 1-2 mg/Kg/day ▪ Omalizumab (Anti-IgE)



Stridor

Definition

It is continuous inspiratory harsh sound due to partial obstruction of the upper airways (Larynx & Trachea). Complete obstruction is fatal

Incidence

Commoner in infants & young children, Why?

Clinical Grading of Stridor

1. Grade I: Stridor with exertion (Crying or exercises)
2. Grade II: Stridor at rest
3. Grade III: Stridor + Retractions (Suprasternal & Supraclavicular)
4. Grade IV: Stridor + Cyanosis

Etiology

A) Acute Stridor

Infectious (= Croup)*	Other Causes
Acute laryngitis	Laryngeal diphtheria
Spasmodic laryngitis	Laryngeal FB
Laryngotracheobronchitis	Laryngospasm
Acute bacterial tracheitis	Laryngeal edema (Post-extubation, allergy)
Acute bacterial epiglottitis	Laryngeal compression (Retropharyngeal abscess)

Important Remarks

- ☒ Infectious croup is much commoner
- ☒ Frequency: Viral > Bacterial
- ☒ Severity: Bacterial > Viral
- ☒ Viral: Parainfluenza, Influenza, Adenovirus, RSV
- ☒ Bacterial Causes: Tracheitis (Staphylococci), Epiglottitis (Hemophilus influenza type b)
- ☒ Laryngotracheobronchitis: Stridor is both inspiratory & expiratory

Infectious (= Croup)	Age	Etiology	Fever	Stridor
Acute laryngitis	1-3 yrs	Viral	Mild	Mild
Spasmodic laryngitis	1-3 yrs	Viral / allergic	Absent	Moderate
Laryngotracheobronchitis	1-3 yrs	Viral	Mild	Moderate
Acute bacterial tracheitis	1-3 yrs	Staph.*	High	Severe
Acute bacterial epiglottitis	3-7 yrs	Hib*	High	Severe

B) Chronic Stridor

Congenital	Acquired
Laryngomalacia	Laryngeal stenosis (Prolonged intubation)
Tracheomalacia	Tracheal stenosis (Prolonged intubation)
Laryngeal web	Laryngeal tumors (Papilloma)
Laryngeal tumor or cyst	Laryngeal paralysis (RLN)
Laryngeal compression (Vascular ring, Goiter)	Laryngeal compression (Tumors)

Management of Stridor

A) Home Management

1. **Observation:** Follow-up of RD
2. **Warm moist environment:** Steamy bathroom (mist therapy)
3. **Drugs**
 - **Steroids:** Dexamethasone should be considered
 - **Antibiotics** are **not** indicated in croup, Why?

The mainstay of Rx is the
Airway Management

Steroids are beneficial
even in mild cases

B) Home Management

Indication of hospital Rx:

- No improvement or deterioration on home treatment
- Stridor grade **III**
- Stridor grade **IV** (It is an indication of endotracheal intubation)
- Infants with grade **II**
- Suspected bacterial etiology (e.g., high fever...)

Components of hospital Rx:

O₂ saturation < 90 % is
an indication of ETT

1. **Close observation**
 - **Clinical:** Vital signs, level of consciousness
 - **Pulse oximeter:** Continuous O₂ saturation monitoring
 - **ABG** may be needed
2. **Minimal disturbances**
 - Keep the child in his preferred position
 - Avoid child anxiety & crying
3. **Oxygen therapy:** *Oxygen should be humidified*
 - Oxygen mask
 - Head box (hood)
 - **Endotracheal intubation** with endotracheal tube (ETT):
 - **Indications:** Cyanosis &/or altered consciousness
 - **Precautions:** Experienced person & using smaller "than usual" ETT
 - **Tracheostomy:** if failed intubation
4. **Helium-oxygen mixture (Heliox):** in severe cases
5. **Warm moist environment:** Nebulizer (± Adrenaline)
6. **IV Fluids**
7. **Drugs**
 - **Antibiotics:** in epiglottitis Ampicillin
 - **Steroids:** Oral / IM Dexamethasone (Or Inhaled steroids; Budesonide)
 - **Nebulized racemic adrenaline:** VC, ↓↓ Edema (0.5 ml in 3 ml saline every 20 min.)
[NB: Racemic epinephrine is not available in Egypt, l-epinephrine is also effective]
 - **Avoid:** Sedatives (↓↓ Conscious level) & bronchodilators (↑↑ O₂ requirement)

Epiglottitis

- ☒ **Etiology:** Hemophilus influenza type b (Now rare, why?), Streptococci
- ☒ **Age:** 3-6 years
- ☒ **C/P:** Fever, toxicity, stridor, drooling, dysphagia, dysphonia, dyspnea, No barking cough
- ☒ **Lateral neck X-rays:** Thumb sign (Swollen epiglottis)
- ☒ **Medical emergency, why?**
- ☒ **Treatment:** ETT is indicated regardless the degree of apparent RD (Usually for 2-3 days)
Antibiotics: Ceftriaxone or (Ampicillin-sulbactam)
- ☒ **Prognosis:** 6% (Without artificial airway) & 1% (With artificial airway)

Chocking (FB Airway obstruction)

Definition

Partial or complete obstruction of the airways (By foods or small objects)

Incidence

Common in late infancy & early childhood, Why?

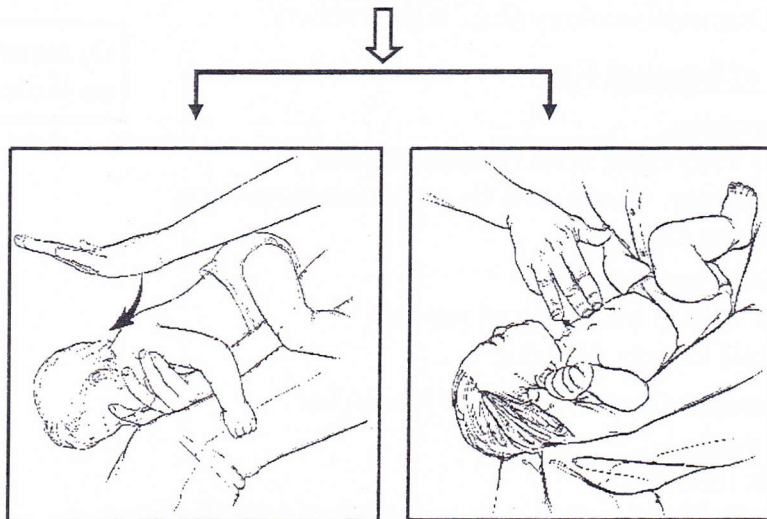
Common in unconscious patients

When to suspect

Sudden onset of cough, choking & cyanosis

Management

Infants



5 Back blows AND 5 Chest thrusts

Children



Abdominal thrust

Respiratory Failure

Definition

- Failure of the respiratory system to maintain adequate levels of O₂ & CO₂
- Most pediatric cardiac arrest begins as respiratory failure

Incidence

- Commoner in infants & young children, Why?
 - **Airways:** Narrower & less supported
 - **Respiratory center:** Immature

Classification (= Types)

	Lung Failure	Pump Failure
Synonyms	▪ Type I respiratory failure ▪ Peripheral respiratory failure	▪ Type II respiratory failure ▪ Central respiratory failure
Basic Defect	Oxygenation defect	Ventilation defect
ABG	Hypoxemia (< 50 mmHg) ± Hypercapnia ± Metabolic acidosis	Hypercapnia (> 60 mmHg) ± Hypoxemia ± Respiratory acidosis
C/P	Respiratory distress (4 grades) 1. Tachypnea & working ala nasi 2. Retraction (Intercostal & succostal) 3. Grunting (forced expiration against closed glottis) 4. Cyanosis	<u>Shallow, irregular, gasping</u> breathing with periods of <u>apnea</u> ± Cyanosis
Therapy	O ₂ therapy ± Ventilation	Ventilation ± O ₂ therapy
Causes	A. Airway disease ▪ Choanal atresia ▪ Laryngomalacia ▪ Epiglottitis ▪ FB inhalation ▪ Vocal cord paralysis B. Lung pathology ▪ Bronhiolitis ▪ Bronhiolitis obliterans ▪ Bronchial asthma ▪ Pneumonia ▪ Pneumothorax ▪ Pleural effusion ▪ Pulmonary edema ▪ Massive lung collapse ▪ Cystic fibrosis ▪ RDS, BPD ▪ Meconium aspiration ▪ ARDS	A. Severe brain pathology ▪ CNS infection ▪ ICH ▪ CNS infarction ▪ Trauma & Tumors ▪ CNS depressants ▪ Central hypoventilation syndrome B. Neuro-muscular apparatus ▪ AHC: Wernicke-Korsakow disease Poliomyelitis ▪ Nerve: Guillain-Barré syndrome ▪ N/M: Myasthenia gravis ▪ Muscle: Myopathy C. Thoracic cage ▪ Kyphoscoliosis ▪ Asphyxiating thoracic dystrophy ▪ Diaphragmatic hernia ▪ Diaphragmatic eventration D. Respiratory muscle fatigue (<u>Type I</u>)

Pathophysiology of RF

- Inspired gas composition
- Alveolar gas composition
- Alveolar ventilation & dead space ventilation
- Diffusion

Respiratory Distress

Definition of RD

- Abnormal respiratory pattern (dyspnea, tachypnea, nasal flaring, retractions, grunting...)
- Respiratory failure can be present without RD (abnormalities of CNS or N/M disease)
- RD can occur without respiratory disease

Causes of RD

A) Pulmonary causes: "Causes of type 1 respiratory failure"

B) Extrapulmonary causes

- ☒ Acute congestive HF: Tachycardia + Tachypnea + Tender hepatomegaly
- ☒ Acute metabolic acidosis: Acidotic breathing (Rapid & deep) e.g., DKA, RTA...
- ☒ Acute severe anemia: Acute hemolytic crisis e.g., G-6-PD deficiency...
- ☒ Sepsis: Due to acidosis & ↑↑ RC

Grades of RD

1. Tachypnea & working ala nasi
2. Retraction
3. Grunting (Forced expiration against closed glottis)
4. Cyanosis:
 - Occurs after failure of all previous compensatory mechanisms
 - Indicates $\text{PaO}_2 < 35 \text{ mmHg}$

Oxygen Therapy

Indications & Dosage of Oxygen

1. Respiratory failure (40-100 %)
2. Cardiovascular failure (40 %)
3. Neurological failure (40 %)

Remember

Normal O_2 concentration
in atmosphere = 21%

Methods of administration

Method	Age group	Comments
Incubator	Neonates	FiO ₂ can be determined by O ₂ analyzer
Head box	Neonates & Infants	
Simple oxygen mask	All ages	FiO ₂ can not be determined
Venturi oxygen mask	All ages	Precise FiO ₂ can be delivered
Nasal prongs (cannula)	All ages	FiO ₂ can not be determined [FiO ₂ =21+ FlowX3]
Ambu bag with (Mask or ETT)	All ages	Delivers 40% O ₂ (If no reservoir) Can deliver 100% O ₂ (If connected to reservoir)
Flow inflating bag (= Anesthesia bag)	All ages	Delivers 100% O ₂ (at all times)
ETT	All ages	Can deliver 100 % O ₂

Monitoring of oxygen therapy

O_2 Sat of $< 85\%$ = ?? hypoxemia

A) Clinical

☑ Color: Pink

☑ Work of breathing: Severity of respiratory distress (Grades)

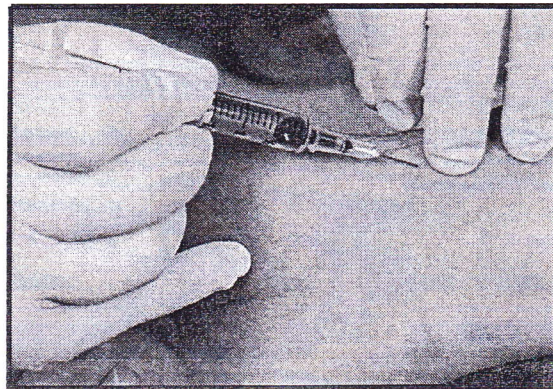
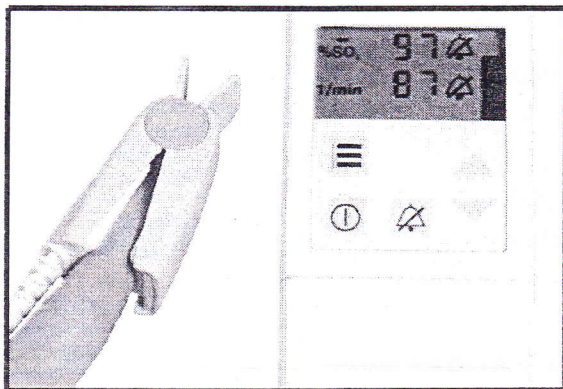
B) Pulse Oximeter: Good response if O_2 Saturation $> 90\%$

C) Arterial blood gases: Good response if O_2 pressure > 80 mmHg

Pulse oximeter Vs ABG

Capnography (= End-tidal CO_2 measurement)

	Pulse Oximeter	ABG
Value	Bed-side (Continuous)	Gold standard for assessment
Parameter	O_2 Saturation	O_2 Tension (& Saturation)
Technique	Rapid non-invasive	Arterial puncture
Unit	Percent	mmHg
Good response	O_2 Saturation $> 90\%$	O_2 Tension > 80 mmHg
Hypoxemia	Mild (90-95 %), Moderate (85-90 %)	< 70 mmHg
Severe hypoxemia	$< 85\%$	< 50 mmHg
Errors (= False results)	▪ Peripheral coldness ▪ Poor peripheral perfusion ▪ Methemoglobinemia ▪ Optical interference	▪ Venous samples ▪ Clots or air in the sample



Complications of oxygen

A) Eye: Retinopathy of prematurity

B) Lungs: Bronchopulmonary dysplasia

C) Oxygen dependence: Difficult weaning

- 100% O_2 is toxic to the lungs in 4 hrs
- 70% O_2 is toxic to the lungs in 4 days
- 40% O_2 is safe for 4 weeks

Indications of Mechanical Ventilation

1. Apnea

2. Hypoxemia: $PaO_2 < 50$ mmHg (in spite of $FiO_2 = 70\% O_2$)

3. Hypoxemia: O_2 Saturation $< 85-90\%$ (in spite of $FiO_2 = 70\% O_2$)

4. Hypercarbia: $PaCO_2 > 60$ mmHg (Hypoventilation)

5. Work of breathing (Respiratory muscle fatigue)

6. Therapeutic hyperventilation ($\uparrow\uparrow$ ICT)

7. Supportive:

- Shock
- After major surgery
- Status epilepticus
- After cardiopulmonary resuscitation (CPR)

Assessment of severity of lung pathology (=O₂ derived pulmonary indices)

1. PaO₂/FiO₂: Normally it is 400-450

- Values < 300: consistent with acute lung injury
- Values < 200: consistent with ARDS

2. Oxygenation index (OI)

- Values between 1-5: Mild lung pathology
- Values between 6-10: Moderate lung pathology
- Values > 20: Severe lung pathology (High mortality)

3. Ventilation index (VI)

$$OI = \frac{MAP \times FiO_2 \times 100}{PaO_2}$$

$$VI = \frac{PIP \times Rate \times PaCO_2}{1000}$$

Approach to a child with RF

Rules

- Initiate investigations in order of priority
- Simultaneous intervention & investigation

Components

A) Assessment

- ☒ History
- ☒ Examination
- ☒ Investigations

B) Respiratory Monitoring

C) Respiratory support

- ☒ Airway patency: Oropharyngeal or nasopharyngeal
- ☒ Oxygen therapy: *See before*
- ☒ Inhaled gases:
 - Helium-oxygen "Heliox" (Helium is less dense than N₂)
 - Nitric oxide: Inhaled pulmonary vasodilator
- ☒ Chest Physiotherapy & Suction
- ☒ PPV
 - CPAP: High-flow nasal cannula (Flow = 4-16 L/min)
 - CPAP: tight-fitting face mask
- ☒ ETT & mechanical ventilation
- ☒ ECMO

D) Specific Therapy

Pneumonia	Anemia
Asthma	Acidosis
Pneumothorax	Stridor
Pleural effusion	RDS
Pulmonary edema	GBS

Endotracheal Intubation

Indications of ETT

1. Persistent hypoxemia or hypercarbia (*despite the above mentioned interventions*)
2. Airway patency (Goiter, micrognathia)
3. Tracheal suction (Meconium aspiration syndrome)
4. Ineffective bag & mask ventilation
5. Drugs e.g., adrenaline...
6. Surfactant therapy (in respiratory distress syndrome)
7. Prolonged ventilation is needed (as in diaphragmatic hernia)

Internal diameter of ETT

$$ID = (Age (yr)/4) + 4$$

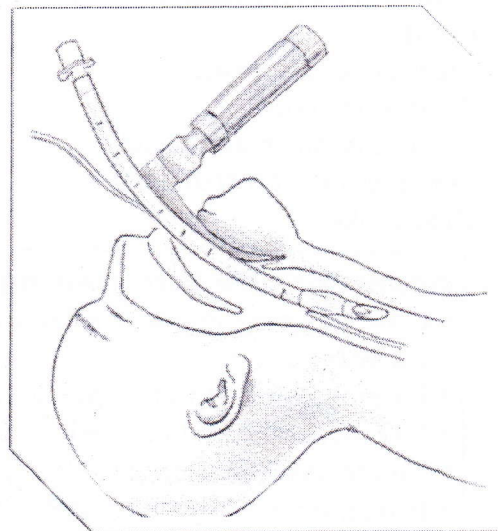
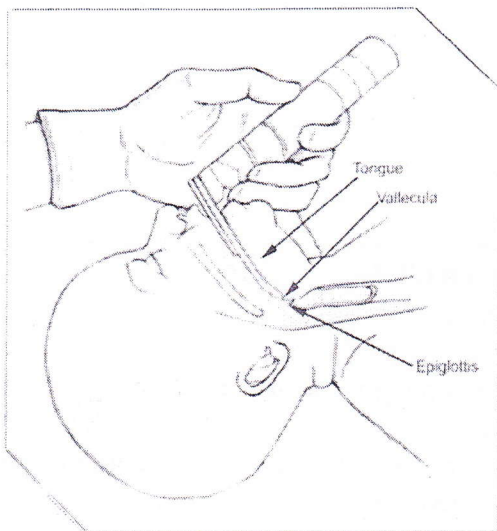
Sedation

- Sedation & paralysis should be considered standard (unless contraindicated)

Category	Drug	Dose	Duration	Comment
Sedatives	Midazolam	0.1 mg/Kg IV	60 min	Amnesia, ↓↓ RC
	Ketamine	1-2 mg/Kg IV	10-15 min	↑↑ HR, ↑↑ BP
Analgesics	Fentanyl	2-5 µg/Kg IV	60 min	↓↓ RC
	Morphine	0.1 mg/Kg IV	120 min	↓↓ RC
N/M blocking agents	Pancuronium	0.1 mg/Kg IV	30 min	↑↑ HR

Check position of ETT

- Seeing the ETT through the vocal cords
- Chest expansion
- Misting of ETT
- No sounds over the epigastrium
- Equal breath sounds
- CO₂ detector or capnography



Sudden Death

(In Infants, Children & Adolescents)

Incidence

- 1-6 per 100.000 per year in children
- It may be traumatic* (Occupational, vehicle or violent deaths) or nontraumatic (Cardiac*)
- 65% of sudden deaths are due to cardiac causes

Causes

1. Sudden infant death syndrome

2. Myocardial disease

- Cardiomyopathy
 - Hypertrophic, Dilated & Restrictive cardiomyopathy
 - Arrhythmogenic right ventricular dysplasia
- Myocarditis

3. Coronary arterial disease

- Anomalous origin
- Anomalous course
- Myocardial infarction
- Kawasaki disease

4. Congenital heart diseases

- Aortic stenosis
- Tetralogy of Fallot
- Transposition of great vessels
- Hypoplastic left heart syndrome
- Duct-dependent CHD
- Mitral valve prolapse
- Eisenmenger syndrome

5. Conduction system (Arrhythmias)

- Long Q-T syndrome
- Pre-excitation syndromes (e.g., WPW syndrome...)
- Heart block
- Commotio cordis ➡

6. Other causes

- Pulmonary embolism
- Pulmonary hypertension
- Child abuse
- Electrolyte disturbances
- Anorexia nervosa
- Cocaine & other stimulants
- Inborn errors of metabolism
- Heat stroke

Commotio cordis

Etiology: Blunt chest trauma

Mechanism: Ventricular fibrillation

Pathology: No cardiac trauma

Prognosis: 90% mortality

Apparent Life threatening events (ALTE)

- **CNS:** AV malformation, seizures, chiari crisis
- **Cardiac:** *as before*
- **Pulmonary:** Aspiration, pulmonary hypertension, pulmonary embolism
- **GIT:-** GE, dehydration, GERD, pancreatitis
- **Endocrine/Metabolic:** CAH, hyperammonemia, GSD type 1, malignant hyperpyrexia
- **Infection:** Meningitis, encephalitis, hepatitis, pyelonephritis, botulism, pertussis, sepsis
- **Trauma:** Child abuse, Munchausen syndrome by proxy
- **Poisoning:** Salicylates, insulin, cocaine, carbon monoxide

Sudden Infant Death Syndrome

Definition

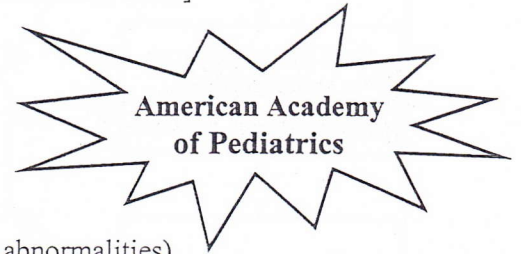
Sudden death of an infant under 1 year of age which remains unexplained after a complete post-mortem examination (DD: congenital anomalies, infection & child abuse)

Incidence

- 0.5 per 1.000 per year (The rate declined **after 1992**, the year in which AAP recommended **Back to Sleep**)
- The 3rd leading cause of infant mortality [8% of all infant deaths in USA]

Pathology

- Petechial hemorrhages
- Pulmonary edema
- Markers of low grade asphyxia
- Higher expression of VEGF in the CSF
- Ventral medulla: Arcuate nucleus hypoplasia (& receptor abnormalities)
- Neuronal & receptor abnormalities



Risk Factors

1. Genetic factors

- Cardiac channelopathies:
 - Sodium cardiac ion channel genes
 - Potassium cardiac ion channel genes
- Serotonin transporter protein
- Genes related to autonomic nervous system development
 - MAO, Tyrosine hydroxylase
 - PHOX2A, PHOX2B
- Genes related to infection & inflammation
 - Complement C4A, Complement C4B, IL-10, IL-6, TNF- α , VEGF
- Genes related to energy production
 - Mitochondrial DNA polymorphisms

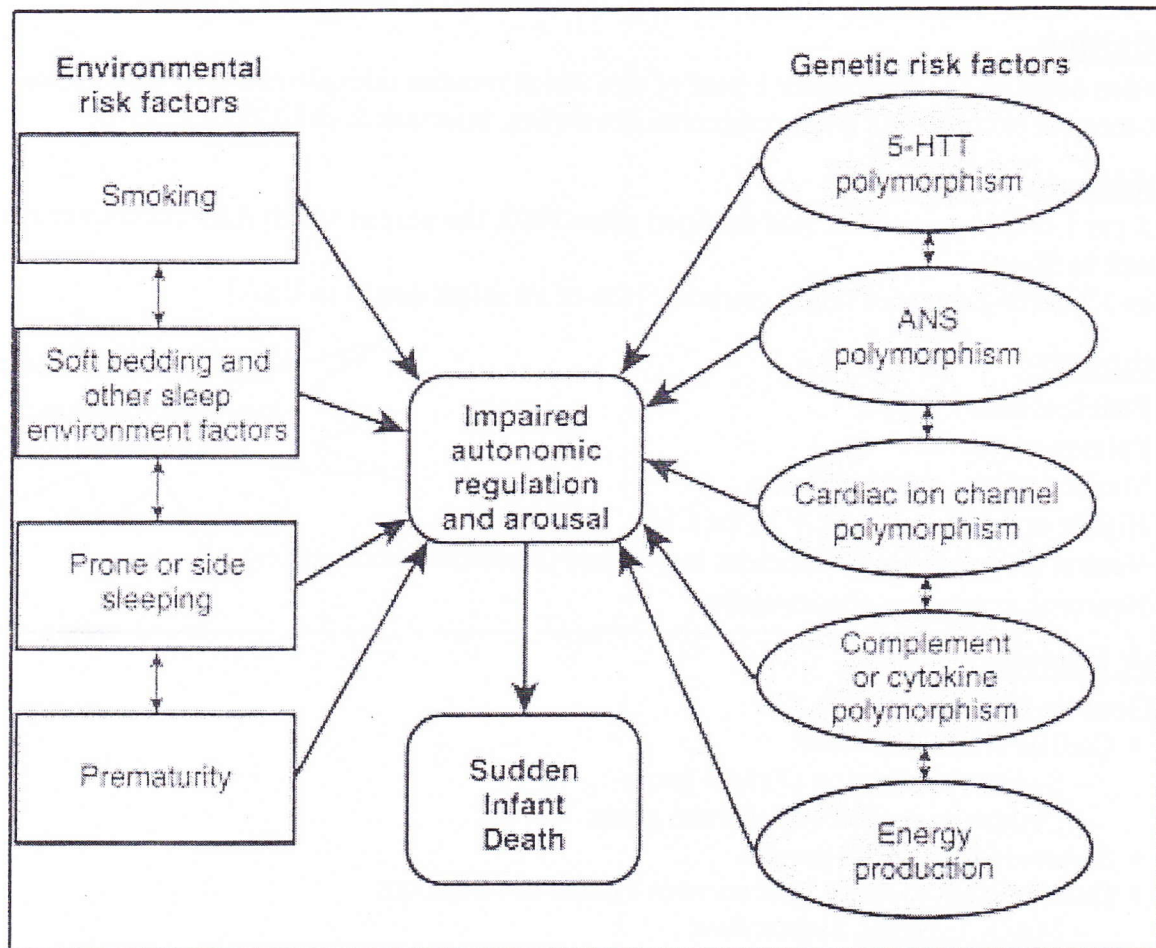
2. Environmental factors

Maternal & Antenatal	Infant
▪ $\downarrow\downarrow$ Age, $\downarrow\downarrow$ Education, $\downarrow\downarrow$ SE level ▪ $\downarrow\downarrow$ Nutrition ▪ Smoking ▪ Alcohol ▪ Drugs (cocaine, heroin) ▪ IU hypoxia ▪ IUGR	▪ Age: Peak 2-4 months, Male sex ▪ Growth failure ▪ Prematurity ▪ Prone & side sleep position ▪ Bed sharing, soft sleeping surfaces ▪ No breastfeeding, No pacifiers ▪ Cold seasons (No central heating)

- Breastfeeding & pacifiers $\downarrow\downarrow$ risk of SIDS
- Supine except in specific situations

Infants at increased risk of SIDS

- A. Infants with ALTE
- B. Siblings of a SIDS victim
- C. Prematurity
- D. Genetic & environmental risk factors



Reducing the Risk of SIDS [As recommended by the AAP]

1. **Supine position** (Prone & side sleeping are not recommended)
2. **Firm mattress** (Avoid soft mattresses)
3. **Sleeping in the parent room** (But in their own crib)
4. **Use blankets with holes**
5. **Avoid soft materials** in the infant's sleep environment (e.g., pillows...)
6. **Avoid bed sharing** (specially if the parents are tired or taking drugs)
7. **Avoid overheating**
8. **Avoid smoking**
9. **No need to use devices** to maintain sleep position
10. **No need to use respiratory, cardiac or O₂ saturation home monitors**
11. Consider offering a **pacifier** at bedtime. The pacifier should be used when placing the infant down for sleep and not be reinserted once it falls out. For breast-fed infants, delay introduction of the pacifier until breast-feeding is well established

Obstructive Sleep Apnea

Definition

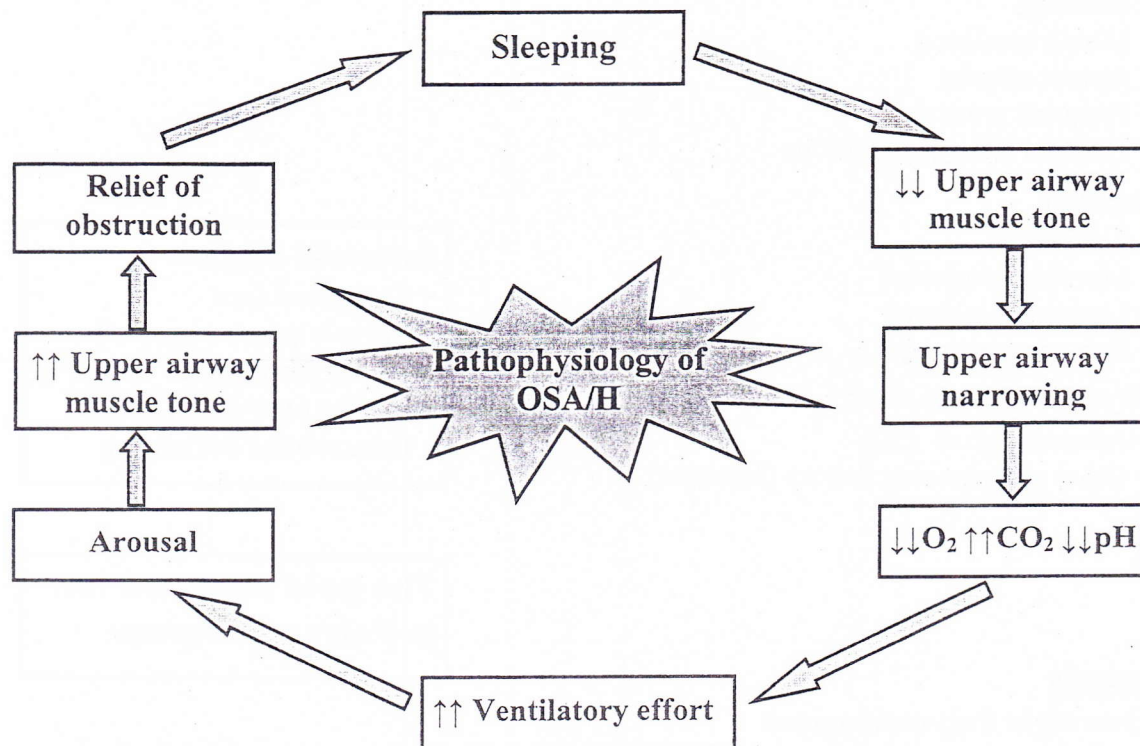
- Repeated episodes of airflow obstruction at the upper airway (Nose & mouth) during sleep
- It is usually due to adenotonsillar hypertrophy
- OSA is part of the spectrum of sleep-disordered breathing (SDB) that also includes:
 - Primary snoring: No ventilatory abnormalities (hypoxia & hypercapnia)
 - Upper airway resistance syndrome: Partial obstruction, snoring & frequent arousal
 - Obstructive hypoventilation (Hypopnea): 50% reduction of airflow
 - Sleep apnea: Complete cessation of airflow; which may be:
 - **Central:** No chest wall movement + No airflow
 - **Obstructive:** Chest wall movement + No airflow
 - **Mixed**

Incidence

- Occasional snoring: 20 %
- Every night snoring: 10 %
- OSA: 1-3 % of pre-school children

Pathophysiology

- Airway obstruction $\Rightarrow$ Hypoxia & hypercapnia $\Rightarrow$ Arousal



Other Effects (= Complications)

- Chronic hypoxia: Polycythemia, growth failure
- Increased pulmonary artery pressure, RV hypertrophy & Cor-pulmonale
- Arrhythmias

Predisposing Factors

Anatomic	Functional
A) Nose - Anterior nasal stenosis - Choanal atresia - Deviated nasal septum - Nasal polyps - Rhinitis B) Mouth, Nasopharynx & Oropharynx - Macroglossia - Cleft palate repair - Adenotonsillar hypertrophy**** C) Craniofacial - Achondroplasia - MPS - Micrognathia - Pierre Robin syndrome	A) REM sleep-related pharyngeal hypotonia B) Neurological causes - Generalized hypotonia (Down...) - Cerebral palsy - Chiari malformation - CNS injury: Hypoxia, trauma, tumors - Autonomic dysfunction C) Drugs - Sedatives: Chloral hydrate... - Narcotics - Anesthetics D) Other causes - Obesity - Dysphagia - Excessive oral secretions

Clinical Picture

A) During Sleep

- Snorting
- Mouth breathing
- Apneic attacks
- Frequent arousals
- Unusual sleeping positions

B) Daytime

- Sleepiness
- Morning headaches
- Learning problems
- Behavioral changes

C) Physical Examination

- Adenoid facies ➡
- Other predisposing factors (*Mention*)

Adenoid Facies

- Elongated face
- Narrow pinched nostrils
- Mouth breathing
- Short upper lip
- Infraorbital darkening

**The gold standard for
is Polysomnogram**

Diagnosis

☒ Overnight Polysomnogram

- **Gold standard** for diagnosis [In practice, Not all children require sleep studies]
- Study of sleep physiologic variables
- Sleep stages, eye movements, muscle tone, airflow, heart rate, O₂ saturation

☒ Other investigations

- Lateral neck X-rays
- CBC
- CXR, ECG & Echocardiography
- ABG, defect?

Treatment

1. Adenotonsillectomy

- It is the **First line of treatment**
- Adenoid may **grow back**, How?
- **↑↑ Risk of postoperative complications with: Obesity, age (< 2 yrs), hypotonia**

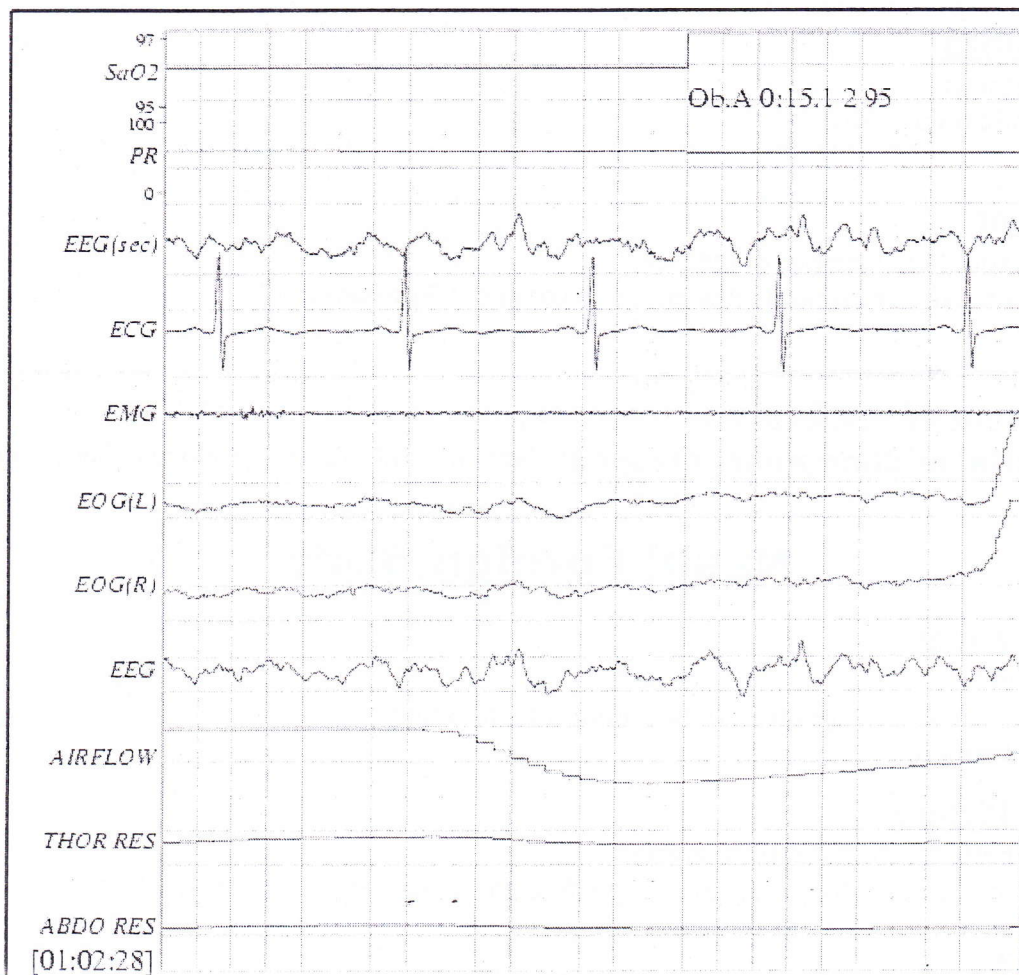
2. Weight reduction

3. Medical therapies

- Nasopharyngeal airway
- Nasal mask CPAP
- Supplemental O₂
- Drugs
 - Topical nasal steroids
 - Antibiotics
 - Nasal decongestants (*Short-term effect*)

4. Surgical therapies

- Craniofacial surgical procedures
 - Mandibular plastic surgery
 - Choanal atresia
 - Cleft palate revision procedures
- Uvulopalatopharyngoplasty
- Correction of deviated nasal septum
- Nasal polypectomy
- Traheostomy



Disorders of the Nose

Introduction

- Most neonates are obligatory nose breathers (Variable)
- Nasal congestion is common in the 1st yr of life
- The internal nasal airway doubles in size in the 1st 6 months of life
- Nose is important in warming & humidification of inspired air
- Nasal mucosa is ciliated
- Nasal secretions contain lysozyme, IgA, lactoferrin, histamine & glycoproteins (Viscid)

Choanal Atresia

Definition & Types

- Septum between the nose & pharynx
- Unilateral or bilateral, bony (90%), membranous (10%) or combined
- CHARGE syndrome: Coloboma, Heart, choanal Atresia, Retardation, Genital, Ear anomalies

Incidence

- 1: 7.000 live births

Clinical Picture

- Unilateral cases may be asymptomatic for long duration (nasal discharge or obstruction)
- Bilateral cases: RD & cyanosis (Improves with crying)

Investigation

- Firm catheter
- Fiberoptic rhinoscopy
- CT

Treatment

- Stabilization: Oral airway & feeding
- Surgical repair: Transnasal (Restenosis is common; Mitomycin C)

Perforation of the Septum: Usually acquired; Syphilis, TB, CPAP

Congenital midline nasal masses: Dermoid, glioma, encephalocele, hemangioma

Nasal Foreign Body

Complications

- Infection
- Local tissue damage (batteries), perforation of the septum
- Nasal obstruction

Clinical Picture

- Beads, batteries, buttons, toys, erasers
- History of FB insertion, nasal discharge, foul nasal odor, epistaxis, obstruction

Treatment

- FB removal: Topical anesthesia, forceps or nasal suction

Epistaxis

Definition

- Bleeding from the nose
- BV of the nose anastomose on anterior part of the septum; Little's area (Kiesselbach's area)

Etiology

A. Local

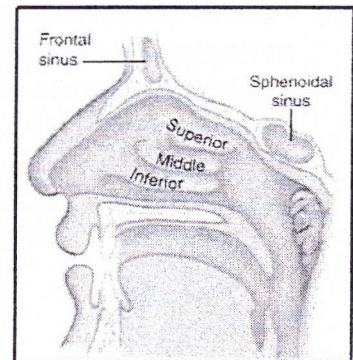
- Idiopathic (Spontaneous)
- Epistaxis digitorum
- FB
- Trauma
- Tumors (juvenile nasal angiofibroma)
- Rhinitis, sinusitis, irritants (GERD, smoking)
- Nasal polyps
- Septal deviation & perforation
- Vascular malformation
- Cocaine, topical steroids

B. General

- Bleeding tendency: platelet, coagulopathy, liver disease,
- Hypertension
- Fever
- Drugs: NSAIDs, aspirin

Treatment

- Compression of the nares with the child in the upright position & the head tilted forward
- Cold compresses
- Local VC (Oxymetazoline; *Afrin*)
- Nasal packing (Anterior or posterior)
- Cautery: Chemical (silver nitrate) or electrical
- Surgical



Nasal Polyps

Definition

- Benign pedunculated edematous nasal mucosa (affecting 0.2-1% of the population)
- They commonly arise from the ethmoidal sinus and occur in the middle meatus
- **Antrochoanal polyp:** arises from the maxillary antrum and can extend to the nasopharynx
- **Samter's triad:** Nasal polyps, aspirin sensitivity & asthma

Etiology

- Cystic fibrosis: Commonest cause in children
- Allergic rhinitis
- Sinusitis

Investigation

- Fiberoptic rhinoscopy, CT

Treatment

- Local or systemic decongestants: Symptomatic
- Steroid spray: Shrinkage of the polyps
- Surgical removal

Common Cold

(Rhinosinusitis- Nasopharyngitis)

Epidemiology

- It is the commonest infection in children (Peak: Fall & Spring)
- Young children have 6-8 colds/year (The incidence ↓↓ with age)
- Cause of recurrence: Many serotypes, superficial infection, antigenic shift & drift
- More in day-care centers

Etiology

- **Viral:** Rhinoviruses, Coronaviruses, RSV, adenovirus, influenza, parainfluenza...
- **Mode of transmission:** Droplet infection & direct contact

Clinical Picture

- **Fever:** Low-grade (may be absent)
- **Sore throat**
- **Nasal symptoms:**
 - Nasal discharge: Initially watery (1st few days), then mucoid
 - Nasal obstruction (Irritability & feeding difficulties)
- **DD:** Allergic rhinitis (Nasal discharge, sneezing, itching, nasal eosinophils)

Complications

1. **Spread of infection**
 - Upper respiratory tract: Otitis media & sinusitis
 - Lower respiratory tract: Acute bronchitis, bronchiolitis, pneumonia
2. Precipitation of asthmatic attack in predisposed children
3. Inappropriate use of antibiotics

Prognosis

- ☒ **Majority:** Benign, self-limited (1 week)
- ☒ **Complications:** Should be considered if



Complications should be suspected if:

- High / persistent fever
- Significant cough
- Persistent nasal discharge > 10 days

Treatment

- Antipyretics
- Oral fluids
- Saline nasal drops ± gentle suction of the nose with a soft bulb "Before feeds"
- Nasal decongestants: Topical (oxymetazoline) or oral (adrenergic agents)
- Antihistamines: ↓↓ Rhinorrhea by 30%

Sinusitis

Etiology

- Viral: Following common cold
- Bacterial: Secondary infection may occur (S. pneumoniae, H. influenza, M. catarrhalis, Staph.)

Clinical Picture

- Nasal symptoms, fever, headache, facial pain, periorbital edema
- Tenderness over the cheek (maxillary sinuses)
- Frontal sinusitis is uncommon, why?

Treatment

- Antibiotics: Amoxicillin or amoxicillin-clavulanate (for 7 days after resolution of symptoms)
- Analgesics

Otitis Media

Incidence

- Acute inflammation of the middle ear
- It is the 2nd most common infection in children with peak incidence 6 months-6 yrs

Etiology (*Bacterial*)

- A) **Common:** Streptococcus pneumoniae, Hemophilus influenza, Moraxella catarrhalis
B) **Others:** Staphylococci, Streptococci, Pseudomonas

Clinical Picture

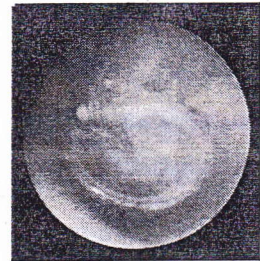
A) Symptoms

- History of previous **common cold** is common
- **Fever**
- **Earache:** Unexplained irritability, crying, pulling on ears
- **Ear discharge:** With drum perforation

B) Signs (Otoscope examination)

- Congested bulging eardrum with loss of normal light reflex
- Discharge (Perforation)

**Ear examination is essential
in every febrile child**



Complications

A) Ear complications

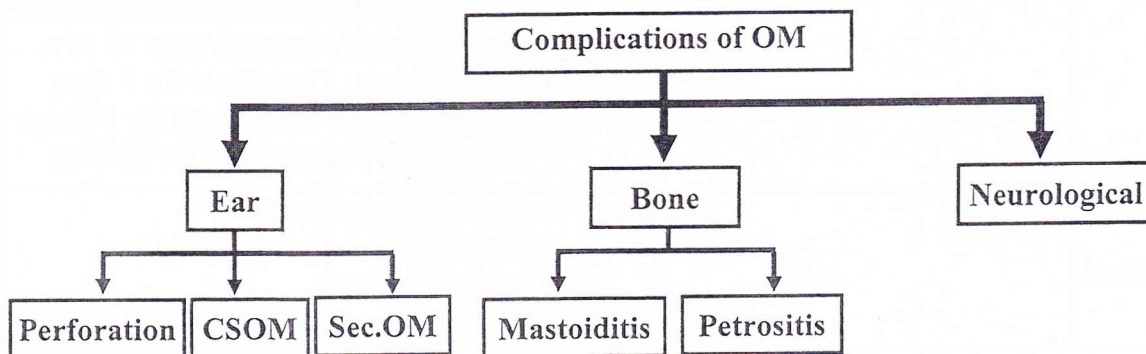
- **Drum perforation**
- **Chronic suppurative OM:** Hearing loss
- **Chronic secretory OM (OM with effusion):**
 - The commonest cause of conductive hearing loss
 - Retracted drum with lost light reflex
 - Rx options: Grommets tubes, adenoidectomy

B) Bone complications

- **Mastoiditis:** Painful swelling behind the ear
- **Petrositis:** Inflammation of the air cells (in the petrous part of the temporal bone)

C) Neurological complications

- **Meningitis, Encephalitis, Brain abscess**
- **Labyrinthitis:** Vestibular ataxia, vertigo
- **Lateral sinus thrombophlebitis**
- **Acute facial palsy**



Treatment

1. Antipyretics & analgesics
2. Antibiotics: Amoxicillin or amoxicillin-Clavulanate for at least 5 days

Pharyngitis

(Tonsillitis)

Epidemiology

- It is a common infection in children > 2 yrs of age (Peak: Fall & Spring)
- Streptococcal pharyngitis is uncommon < 2-3 yrs

Etiology

A) **Viral***: Adenoviruses, coronaviruses, rhinoviruses, RSV, Coxsackie, EB virus

B) **Bacterial**: Group A β -hemolytic streptococci* [**M protein** is the major virulent factor]

Clinical Picture

	Viral Pharyngitis	Streptococcal Pharyngitis (Tonsillitis)
Fever	Mild-moderate	High-abrupt
Sore throat	Moderate	Severe (Difficult swallowing)
Other Symptoms	Diarrhea	Headache, abdominal pain, vomiting Rash in scarlet fever
Throat Ex.	▪ Mild erythema of tonsils & pillars ▪ Small ulcers on the soft palate or posterior pharyngeal wall may occur	▪ Diffuse redness of tonsils & pillars ▪ Follicular exudation (Follicular tonsillitis) ▪ Membrane formation (Membranous tonsillitis)
LN	-	Enlarged tender anterior cervical LN
Cough Rhinitis Conjunctivitis Hoarseness	Common	Rare
Lab	➤ Viral culture? ➤ PCR?	➤ Throat culture ➤ Rapid antigen test
Treatment	No specific Rx	➤ Oral Penicillin V: for full 10 days; (Even with early improvement) ➤ IM Benzathine penicillin: 600,000-1,200,000 IU (Sensitivity test is required) ➤ Oral amoxicillin for 10 days ➤ Cephalosporin (Cephalexin) For patients allergic to penicillin ➤ Erythromycin: 40mg/Kg/d for 10 days ➤ Azithromycin: 12mg/Kg/d for 5 days ➤ Clarithromycin: 15mg/Kg/d for 10 days ➤ Clindamycin: 20mg/Kg/d for 10 days
Complications	OM	➤ Retropharyngeal abscess ➤ Peritonsillar abscess (quinsy) ➤ Rheumatic fever ➤ Post-Streptococcal GN

Indications of tonsillectomy: (Controversial)

1. Recurrence $\geq$ 3-5/yr
2. Peritonsillar abscess (Quinsy)
3. Obstructive sleep apnea

NB: Large tonsils alone is NOT an indication

Multivalent vaccine is under development

Acute Bronchitis

Incidence

It is the most common cause of acute cough in children

Classification

A) **Nonspecific Bronchitis:** Mostly viral

B) **Specific Bronchitis:** With measles, pertussis, diphtheria, scarlet fever

Etiology

A) **Viral*:** Following nasopharyngitis (Rhinoviruses...)

B) **Bacterial:** Pneumococci, Staphylococci, Hemophilus influenza

Clinical Picture

- History of previous **nasopharyngitis** is common (3-4 days before the onset)
- **Cough**, chest pain
- **Fever:** High fever suggests bacterial etiology
- **Vomiting:** Gastric irritation, why?
- **Duration:** 2 weeks

Stages (3 stages)

	Early stage	Productive stage	Convalescent stage
Cough	Dry Severe Metallic (Brassy) Spasmodic (with tracheitis)	Productive Less severe Rattling	Less frequent Less severe May be prolonged (2 wks)
Chest Ex.	Normal	Expiratory rhonchi Coarse crepitations	Chest signs disappear gradually

Treatment

1. Cough medicines

- Expectorants & mucolytics may be prescribed to liquefy viscid secretion
- Drinking water is the best expectorant
- Cough suppressants should be avoided

2. Antibiotics:

- When bacterial bronchitis is suspected
- Amoxicillin for 5 days

Bacterial bronchitis is suspected if:

- High fever
- Sick look
- Prolonged cough
- Purulent sputum

DD: Causes of cough

Acute Bronchiolitis

Incidence

- It is the most common cause of RD & acute wheezing in infants resulting from inflammatory obstruction of the small airways (bronchioles)
- Age: 1st 2 yrs of life (Infancy), with peak incidence at 3-6 months (Winter)

Etiology

- A) **Viral:** Respiratory syncytial virus (> 50% of cases), parainfluenza, adenoviruses
- B) **Others:** Mycoplasma

Pathophysiology

- Viral infection → Inflammation of the small airways → Obstruction (Edema & mucus)
- Obstruction is more during expiration → Expiratory wheezes
- Ventilation / Perfusion mismatch

Clinical Picture

A) Symptoms

- History of contact with a family member with minor respiratory illness
- Development of **nasopharyngitis** (3-4 days before the onset)
- **Cough, wheezing, dyspnea, RD, irritability** following URT symptoms
- Apnea in infants < 6 month
- Risk factors for severe disease: age < 12 wks, GA < 35wks, BPD, CHD

B) Signs

- **RD** (Grades 1-4)
- **Expiratory wheezes**

Investigations

- **CBC:** Normal
- **CXR:** Hyperinflation
- **Nasopharyngeal wash:** for RSV antigen detection (Sensitivity = 90%)

Prevention

- Palivizumab (IM, monthly): Monoclonal antibody to RSV [BPD & preterm infants]
- RSV IVIG

Treatment

A) **Mild RD:** Can be managed at **home**

1. Careful feeding (To avoid aspiration)
2. Close observation (Degree of RD)

B) **Moderate to Severe RD &/or risk factors:** Should be **hospitalized**

1. Oxygen therapy
2. Proper hydration: IV fluid
3. Nebulized β -agonists: Salbutamol [Nebulized hypertonic saline may have some benefit]
4. Steroids (Controversial)
5. Antibiotics are **not** indicated except if there is...
6. Antiviral agents (as ribavirin) may be used in infants with CHD or chronic lung disease (Not indicated in otherwise healthy infants)
7. Mechanical ventilation may be needed in severe cases (1% of cases)

Prognosis

- **Recovery:** Usually occurs in 7-10 days
- **Mortality:** < 1%

Cystic Fibrosis

Definition

- Inherited multisystem disorder characterized mainly by:
 - Recurrent **chest** infections $\Rightarrow$ Chronic lung damage (Bronchiectasis)
 - GIT malabsorption $\Rightarrow$ Failure to thrive

Molecular Genetics & Epidemiology

- Autosomal recessive (CFTR gene is located on chromosome 7q)
- It is due to a defect in CFTR (CF transmembrane regulator) protein
- There are >1500 mutations (grouped into 5 Classes of mutations)
- Loss of phenylalanine at position 508 ($\Delta F508$) is the commonest ($\approx 75\%$)
- **Modifier** gene polymorphisms lead to variation in disease severity (TGF- β 1 gene)
- "Mild" mutation of CFTR can cause isolated isolated congenital bilateral absence of the vas
- Incidence: 1/ 3,500 live births
- Highest prevalence in Northern Europeans [Carrier rate = 1:25]

CFTR Structure & Function

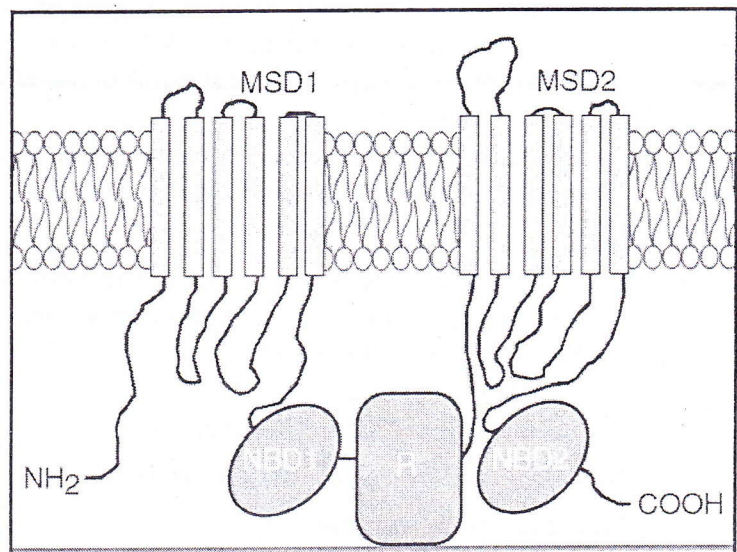
- CFTR is a glycoprotein (1480 amino acids) arranged in 5 domains
- It acts as Cl channel which is activated by phosphorylation of the R domain (cAMP-mediated)
- CFTR is located at the apical compartment of **epithelial** cells in respiratory, GIT, GU, sweat & salivary glands

CFTR Defect:

- **Respiratory, GIT:** Defective Cl secretion & $\uparrow\uparrow$ reabsorption of Na & water
Viscid secretion
- **Sweat glands:** [The main function of sweat glands to absorb not to secrete Cl]
Elevation of sweat Na & Cl (**Basis of Sweat chloride test**)
- **CFTR dysfunction:** Chloride is trapped inside the cells in the airway and outside in the skin

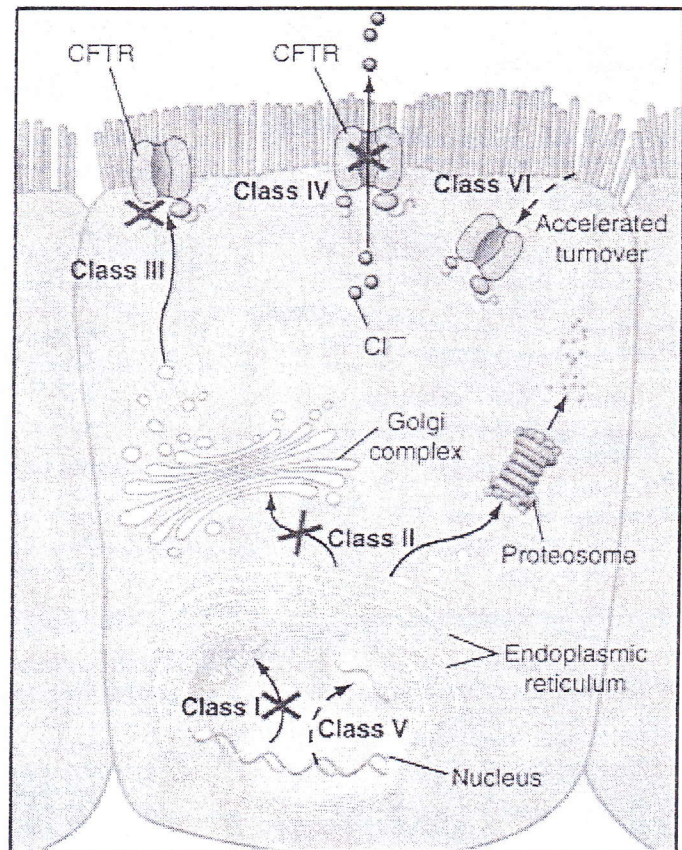
Domains of CFTR (5):

- 2 membrane-spanning domains
- 2 nucleotide-binding domains
- 1 regulatory domain



Categories of CFTR mutations:

- Class I: Absent protein synthesis
- Class II: Defective protein maturation
- Class III: Defective regulation
- Class IV: Defective Cl⁻ conductance
- Class V: Defective transcription
- Class VI: Accelerated turnover



Pathogenesis & Pathology

☒ Respiratory system

- Viscid secretion ($\downarrow\downarrow$ H₂O & $\downarrow\downarrow$ HCO₃), acidified surface liquid
- Difficult clearance of secretion by mucocilliary mechanisms
- Airway colonization with Staph. aureus, H. influenza, Pseudomonas, B. cepacia
- Chronic infections (nutritional deficit is a contributing factor)
- Inflammatory response (mediators) in the wall
- Airway obstruction
- Bronchitis, bronchiolitis, bronchiectasis, emphysema, pneumothorax, fibrosis
- Nose & sinuses: Epithelial hyperplasia, nasal polyps & sinusitis

☒ GIT

- Viscid pancreatic secretion (exocrine dysfunction) with later endocrinal dysfunction
- Pancreatic pathology: eosinophilic material & fibrosis
- Salivary gland obstruction
- GERD (Chronic cough)
- Liver dysfunction & biliary cirrhosis

☒ Sweat gland: Positive sweat chloride test

☒ Genitourinary system

- Males: Obliteration or atresia of the epididymis, vas deferens & seminal vesicles
- Females: Cervical glands are distended with mucus, endocervicitis

Clinical Picture

A) Respiratory Tract (*Main determinant of morbidity & mortality*)

- Cough is the most constant symptom (1st dry but then productive purulent)
- Recurrent chest infections: Staph. aureus, H. influenza, Pseudomonas, B. cepacia
- Bronchiolitis (wheezes)
- Examination: Clubbing & chest signs (shape, air entry, wheezes, crepitations)
- Complications: atelectasis, hemoptysis, pneumothorax, fibrosis, cor pulmonale
- Nasal polyps (15-20%) & chronic sinusitis

B) Cardiovascular system

- RV hypertrophy & failure (= Cor pulmonale) is a late event

C) Pancreas

- Exocrinal dysfunction: Maldigestion & malabsorption $\Rightarrow$ FTT (Bulky greasy stools)
- Endocrinal dysfunction: Diabetes mellitus (2nd decade) $\Rightarrow$ Polyuria & polydipsia
- Acute pancreatitis

D) Intestinal Tract

- Delayed passage of meconium
- Meconium ileus in 15-20 % of neonates with CF
- Meconium peritonitis
- Meconium plug syndrome
- Distal intestinal obstruction syndrome (DIOS)
- Intussception, fecal impaction, rectal prolapse
- Vitamin deficiency $\Rightarrow$

Vit A: Keratomalacia, xerophthalmia, blindness, bitot spots, infections Thick skin
Vit D: Rickets & osteoporosis
Vit K: Bleeding
Vit E: Hemolytic anemia, PN, myopathy

E) Hepatobiliary

- Hepatomegaly & liver dysfunction
- Biliary cirrhosis & portal hypertension
- Biliary stones (Cholelithiasis)

F) Sweat glands

- Hyponatremic dehydration (Salt loss)
- Hypochloremic alkalosis
- Salty taste of sweat (?kissing)

G) Genitourinary Tract

- Male infertility is common (> 95%): Due to obstruction of the vas deferens in utero resulting in atresia or absence at birth [Azoospermia]
- Female fertility is usually maintained: Pregnancy outcome is related to general health & nutritional status of the mother

Diagnostic Criteria of CF

Clinical

- Typical clinical features
OR
- Sibling with CF
OR
- Positive neonatal screening

PLUS

Laboratory

- Two positive sweat Cl test
OR
- Two CF gene mutations
OR
- Abnormal nasal potential difference

Investigations

A) Laboratory

1. Sweat chloride test

- Value: Gold standard for diagnosis (**Some mutations have normal sweat chloride**)
- Technique: 100 mg sweat is needed (using pilocarpine iontophoresis)
- Results: $Cl > 60$ (Positive), 40-60 (borderline), < 40 mEq/L (normal)
- Should be repeated, why?
- False negative test: Malnutrition, dilution, certain mutation
- False positive test:
 - Autonomic dysfunction
 - Addison disease & CAH
 - Anorexia nervosa
 - Hypoproteinemia
 - Malnutrition
 - Dehydration
 - Nephrogenic DI
 - MPS & GSD type 1
 - Hypothyroidism
 - Hypoparathyroidism (familial)
 - Eczema
 - Ectodermal dysplasia

2. Immune-reactive trypsin (IRT): $\uparrow\uparrow$ (Used < 3 months of age)

3. Nasal potential difference: $\uparrow\uparrow$

4. Neonatal screening: By IRT & limited DNA testing

5. DNA testing:

- Diagnosis of CF: For the common mutations ($\Delta F508$ & up to 30 other mutations)
 - Prenatal diagnosis
 - Carrier detection
- } mutational analysis

6. CBC, ABG, electrolytes & Liver function tests

7. Sputum culture (Fasting gastric aspirate, lower pharyngeal swab, bronchoscopy)

8. Pulmonary function tests: Obstructive changes (Late: restrictive)

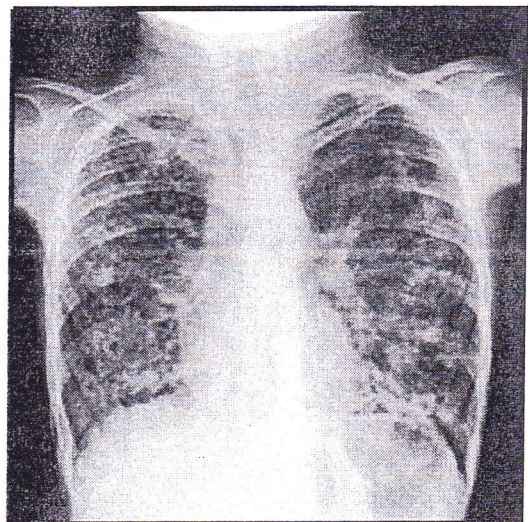
9. Pancreatic function

- Diagnosis of fat malabsorption: 3-day stool collection or serum carotene
- Duodenal aspirate
- Stool elastase is $\downarrow\downarrow$
- OGTT

B) Imaging

1. CXR, CT chest

- Hyperinflation
- Infiltrations & opacities
- Bronchial thickening
- Bronchiectasis
- Dilated pulmonary artery
- CT is sensitive to early bronchiectasis

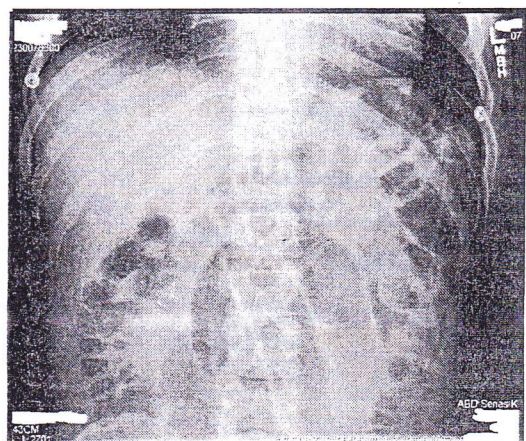


2. CT Sinuses

- Failure of frontal sinuses development
- Panopacification of sinuses

3. Abdominal X-rays

- Dilated loops with air-fluid levels
- Ground-glass material in the lower abdomen
- Peritoneal or scrotal calcification



C) Invasive

1. Bronchoscopy

2. GIT endoscopy

Treatment

A) Pulmonary therapy

1. Inhalation therapy (Inhaler, spacer, nebulizer)

- Saline & hypertonic saline
- Bronchodilators
- Steroids
- Human recombinant DNase: Single daily dose (Improves pulmonary function)

2. Airway clearance therapy

- Chest physiotherapy: Chest percussion & postural drainage
- Expectorants: Iodides (?efficacy)

3. Antibiotics: Oral, Aerosolized or IV. Organisms??

- NB: Infection with mycoplasma & chlamydia has been documented. So, macrolides may be used empirically for flare of symptoms

	Organisms	Agents
Oral	Staph. aureus	Dicloxacillin, Cephalexin, Amoxicillin-clavulanate
	H. influenza	Amoxicillin
	Pseudomonas	Ciprofloxacin
	Burkholderia cepacia	SMZ-TMP
Aerosol		Tobramycin Gentamycin
IV	Staph. aureus	Nafcillin Vancomycin
	Pseudomonas	Tobramycin Amikacin Piperacillin Ceftazidime
	Burkholderia cepacia	Chloramphenicol

4. Bronchodilators

- B-adrenergic agonists, ipratropium, cromolyn sodium

5. Anti-inflammatory

- Steroids: in severe reactive airway disease & allergic bronchopulmonary aspergillosis
- NSAIDs (Ibuprofen)

6. Endoscopy & lavage

7. Rx of pulmonary complications

- Atelectasis
- Hemoptysis: Antibiotics, physiotherapy, vitamin K
- Pneumothorax
- Respiratory failure
- Rt sided heart failure: Diuretics
- Allergic aspergillosis: oral steroids, oral anti-fungal Rx
- Non-tuberculous mycobacterial infection

8. Lung transplantation

9. Nasal polyps

- Local or systemic decongestants: Symptomatic
- Steroid spray: Shrinkage of the polyps
- Surgical removal

10. Rhinosinusitis: Antibiotics

B) Nutritional therapy

1. **Diet:** High calorie (130 Kcal/Kg/day), Low-fat diet (Elemental formula)
2. **Vitamin supplementation**
3. **Mineral supplementation:** Zinc, iron...
4. **Pancreatic enzyme replacement:** ↑↑ doses is linked to colonic stricture
5. **Recombinant GH therapy**
6. **Salt loss**
 - Sweat salt loss specially in hot weather
 - Avoid overdressing
 - Free access to salt & water

C) GIT

1. **Portal hypertension & liver cell failure**
2. **Meconium ileus:** Fluid intake, laxatives, gastrografin enema
3. **DIOS (Meconium equivalent)**
 - Polyethylene glycol (*Non-absorbed laxative*)
 - Large volume bowel lavage (Oral or NGT)
 - Complete obstruction: Enema
4. **Intussusception, volvulus**
5. **GERD**
6. **Pancreatitis**
7. **Diabetes mellitus**

D) Other lines: Gene therapy

Prognosis

- Life-limiting disease (But survival has dramatically improved)
- Males have better survival
- Activities should not be restricted, regular physical exercise should be encouraged

Complications of Treatment

Complication	Agent
GITl bleeding	
Hyperglycemia	
Growth retardation	
Renal dysfunction:	
Tubular	
Interstitial nephritis	
Hearing loss, vestibular dysfunction	
Peripheral neuropathy or optic atrophy	
Hypomagnesemia	
Hyperuricemia, colonic stricture	
Goiter	
Gynecomastia	
Enamel hypoplasia or staining	

Primary Ciliary Dyskinesia

(Immotile Cilia Syndrome)

Definition

- Inherited disorder characterized by impaired ciliary function

Molecular Genetics & Epidemiology

- Autosomal recessive (Rare cases are AD & XL)
- Genes involved in PCD include DNAI1 (9p) & DNAH5 (5p)
- Incidence: 1/ 20,000 live births

Normal Ciliary Ultrastructure and Function

- Cilia are hair-like organelles that move fluids, mucus, and inhaled particulates
- The epithelium in the nasopharynx, middle ear, sinuses & larger airways is ciliated
- A mature ciliated epithelial cell has ≈ 200 uniform motile cilia
- Each cilium is a complex composed of ≈ 250 proteins
- Dynein (ATPase): arranged in outer & inner dynein arm
- Hydrolysis of ATP is the source of energy (ciliary movement)

Types of Cilia:

- **Motile cilia:** Respiratory system
- **Primary cilia:** Immotile (Nephron, retina, bile ducts, hypothalamus)
- **Nodal cilia:** Only during embryonic development, rotational movement, body sidedness

Structural changes in PCD:

- Shortening or absence of dynein arms is the most common (90%)
- Other anomalies: Microtubular transposition, ciliary agenesis, radial spoke & nexin link defects

Clinical Picture

A) Respiratory Tract

- Neonatal RD
- Chronic cough
- Recurrent pneumonia
- Bronchiectasis
- Airway colonization with H. influenza, Staph. aureus, S. pneumoniae, Pseudomonas
- Ear: Chronic otitis media & hearing loss
- Rhinitis, nasal polyps & chronic sinusitis
- Examination: Clubbing & chest signs (shape, air entry, wheezes, crepitations)
- Complications: atelectasis, hemoptysis, pneumothorax, fibrosis, cor pulmonale
- The average age of diagnosis is > 4 yr (*High index of suspicion is needed*)

25% of patients with situs inversus totalis have PCD

B) Genitourinary Tract

- Male: Immotile sperms (Typical but not always present)
- Female: Fallopian tubes

C) Orientation (Laterality) defects

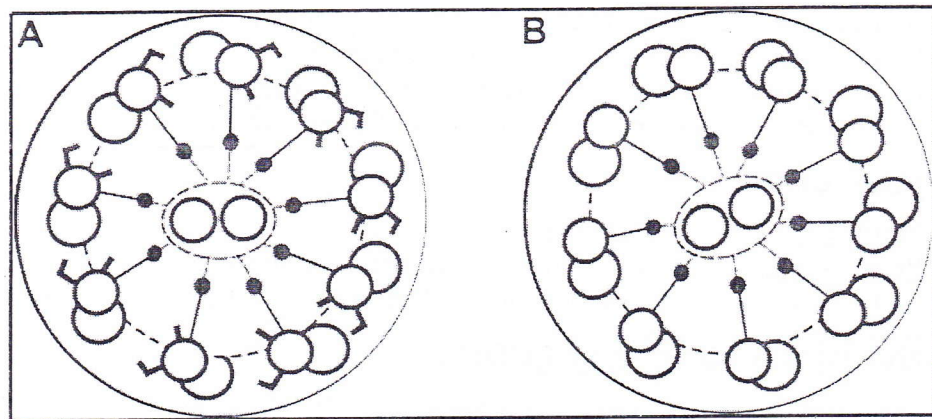
- 50% of cases have situs inversus totalis
- **Kartagener triad:** situs inversus totalis, chronic sinusitis & bronchiectasis
- Heterotaxy (asplenia, polysplenia, CHD)

D) CNS

- Hydrocephaly, why?
- Retinitis pigmentosa

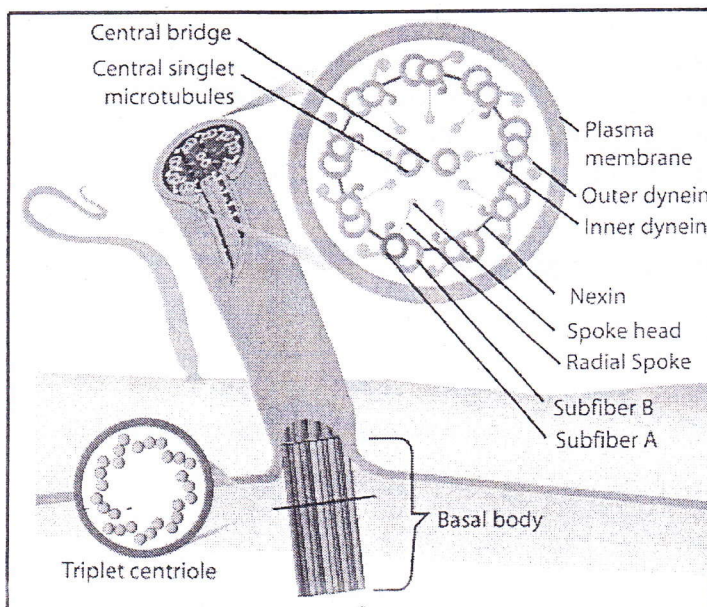
Investigations

- CXR & CT chest
- CT Sinuses
- Electron microscopy:
 - The **gold standard** to assess ciliary ultrastructural defects
 - Nasal epithelium or bronchial brushing can provide an adequate specimen
 - Ciliary defects can be acquired, how?
 - Normal ultrastructure does not exclude PCD
- Qualitative tests to assess ciliary function: Saccharin test
- Exhaled NO concentration ($\downarrow\downarrow$ in PCD)
- Ciliary beat frequency measurements
- High-resolution, high-speed, digital imaging of ciliary motion
- Genetic testing



Treatment (No specific Rx)

- Airway clearance therapy
- Activities should not be restricted, regular physical exercise should be encouraged
- Bronchodilators
- Antibiotics
- Sinus & ear



Ciliopathies:

- PCD
- Bardet-Biedl syndrome
- Polycystic kidney and liver disease
- Nephronophthisis
- Joubert syndrome
- Senior-Loken syndrome
- Alstrom syndrome
- Meckel-Gruber syndrome
- Asphyxiating thoracic dystrophy
- Some forms of retinal degeneration
- Some forms of hydrocephaly

Emphysema & Overinflation

Definitions

- **Emphysema:** Distension of the air spaces with destruction of the alveolar walls (Irreversible)
- **Overinflation:** Distension of the air spaces $\pm$ destruction of the alveolar walls (Reversible)
- **Compensatory overinflation:** Occurs in normal lung tissue 2ry to pneumonia, collapse...
- **SC emphysema:** Air in the SC tissue (Pneumothorax, tracheostomy, fracture orbit, asthma, trauma, toracocentesis...). Crepitus, self-limited condition, No specific Rx
- **Obstructive overinflation:** 2ry to partial obstruction (Ball-valve mechanism) by FB, thick mucus, LN, tumors, TB...

A. Localized Obstructive Overinflation

a. Unilateral pulmonary emphysema (Macleod syndrome):

- Postinfectious; following pneumonia or obliterative bronchiolitis (?Adenovirus)
- Pathogenesis: Arrest of alveolar growth & hypoplasia of pulmonary vessels
- Radiology: Small & hyperlucent lung with overexpansion of the contralateral lung

b. Congenital lobar emphysema

- **Etiology:**
 - Deficient bronchial cartilage
 - Obstruction (aberrant vessel, mucosal flap)
 - Familial cases have been reported
- **Pathology:**
 - Left upper lobe is the commonest to be affected
 - Collapse of the ipsilateral normal lung & compression on the opposite lung
 - Mediastinal shift & transmediastinal herniation
- **Clinical Picture:**
 - Variable degrees of RD
- **Investigations:**
 - CXR, CT, bronchoscopy, V/Q scan
 - MRA, why?
- **Treatment:**
 - Medical Rx: Selective intubation of the unaffected lung, bronchoscopy
 - Surgical excision

B. Generalized Obstructive Overinflation

(Diffuse involvement of the bronchioles)

- **Etiology:**
 - Asthma, cystic fibrosis, bronchiolitis
- **Pathology:**
 - interstitial emphysema, pneumomediastinum & pneumothorax
- **Investigations:**
 - CXR: Hypertranslucency of both lungs
 - Transverse ribs with wide spacing
 - Low flat diaphragm
 - ↑↑ AP diameter
 - Ribbon-shaped heart
 - Pneumatocoeles ➡



α 1-Antitrypsin Deficiency

(& Emphysema)

Definition

- Inherited disorder characterized by:
 - Important cause of emphysema in adults (3rd & 4th decades)
 - Important cause of liver disease in children

Molecular Genetics & Epidemiology

- Autosomal recessive "Codominant" (A1AT gene is located on chromosome 14q)
- Incidence: 1/ 5,000 live births (*Northern Europeans**)

A1AT Function

- A1AT is a protease inhibitor (Pi) synthesized in the liver (> 100 variants)
- It protects tissues from enzymes of inflammatory cells (especially neutrophil elastase)
- Cigarette smoke can lead to oxidation of methionine 358 of A1AT (essential for binding elastase)
- The normal α 1-AT is PiM, mutant protein is not produced (null), or abnormal (PiZ & others)
- Serum level for Enzyme phenotypes:
 - Pi MM: 100%
 - Pi MZ: 60%
 - Pi ZZ: 15%
 - Pi nullnull: 0% (No liver disease)

Effects of A1AT deficiency:

- Break down of elastin (Elasticity of the lungs), resulting in emphysema in adults and cirrhosis in adults & children

Clinical Picture

A) Respiratory Tract

- Emphysema
- Smoking greatly increases the risk of emphysema
- Chronic respiratory symptoms

No or little detectable pulmonary disease during childhood

B) Hepatic

- Neonatal cholestasis, hepatomegaly, splenomegaly

C) Skin: Vasculitis

Investigations

- Enzyme level: Normal: 150-350 mg/dL
- Serum electrophoresis for enzyme phenotype
- PCR for genotype
- CXR & CT chest: Emphysema
- PFTs: usually normal in children, but it can show airflow obstruction particularly in adolescents who smoke
- Neonatal screening

Treatment

- IV enzyme replacement from pooled human plasma (A level of 80 mg/dL is protective)
- Recombinant A1AT: under trial
- Inhalation of the plasma-derived product is under evaluation
- Lung transplantation for end-stage disease

Congenital Anomalies of the Lung

Pulmonary agenesis & Aplasia

Definition

- Pulmonary agenesis: Complete absence of a lung (No bronchial stump or carina) [AR disease]
- Bilateral pulmonary agenesis is incompatible with life
- Pulmonary aplasia: There is bronchial stump & carina

Incidence

- Pulmonary agenesis: 1:10.000 live births
- Rt lung agenesis has higher mortality than the Lt
- May be associated with other congenital anomalies (VACTERL)

Clinical Picture

- Manifestations are usually related to central airway complications; compression, stenosis...
- Scoliosis & thoracic asymmetry, why?

Investigations

- CXR (Mediastinal shift to the same side), CT is diagnostic

Treatment Supportive

Pulmonary Hypoplasia

Definition

- ↓↓ Number of alveoli & airway generations

Etiology

- Primary
- Secondary: Oligohydramnios (*causes*), diaphragmatic hernia, CCAM, thoracic dystrophy, pleural effusion (hydrops)

Clinical Picture

- RD, PPHN

Treatment Supportive: RD, PPHN

Lung Hernia

Definition

- Herniation of the lung beyond its normal boundaries; usually **cervical**
- Other sites include: Parasternal, paravertebral, intercostal

Etiology

- Congenital
- Acquired: Trauma, chronic cough, weak neck muscles (Trapezius, scalene muscles)

Clinical Picture

- Cervical hernia: Neck soft swelling that appear with straining
- Other sites

Treatment

- Rx of the cause
- Reassurance. Surgical Rx for cosmetic reasons may be needed

Congenital Cystic Adenomatoid Malformation

Definition

- Hamartomatous or dysplastic lung tissue mixed with normal lung confined to one lobe
- Cysts are very common (may contain cartilage)

Types

	Type 1	Type 2	Type 3
Incidence	50	40	10
Histology	Macrocytic	Microcystic	No cysts
Prognosis	Good	Bad	Poorest

Clinical Picture

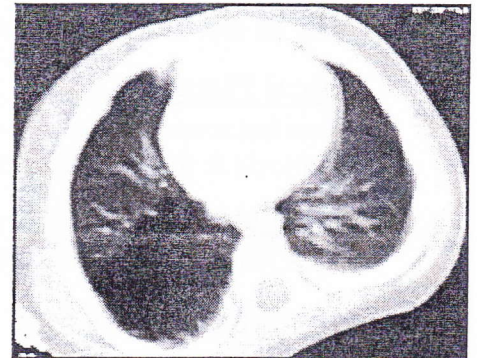
- RD, respiratory infection, pneumothorax
- Pulmonary hypoplasia may develop, why?
- May be asymptomatic for years

Investigations

- CXR & CT (Cystic mass, DD: Diaphragmatic hernia)

Treatment

- Antenatal Rx: Controversial
- Surgical Rx for symptomatic patients
- Surgical Rx for asymptomatic patients by 1 yr (Risk of carcinoma or sarcoma)



Pulmonary Sequestration

Definition

- Congenital anomaly in which part of the lung (sequestered tissue) is not connected to the normal bronchial tree & receives its arterial supply from the systemic arteries (not pulmonary)
- It may be intralobar or extralobar

Types

	Intralobar	Extralobar
Incidence	75%	25%
Onset	Adolescence	Infancy
Site	Lower lobe	Left lung (90%)
Visceral pleura	Covered with the same pleura	Has its own pleura
Sex	M = F	M > F (80%)
Venous drainage	Pulmonary veins (LA)	IVC (RA)
Infection	Common	Rare
Associations	Rare	CCAM, CDH, pulmonary hypoplasia CHD, vertebral & colonic duplication
Picture		

Investigations CXR, CT, MRA

Treatment Surgical excision

Bronchogenic Cysts

Definition

- Cysts due to defective growth of the embryologic lung bud
- Lined with ciliated epithelium
- Commonly on the Rt side near midline (trachea, esophagus, carina)

Clinical Picture

- Compression: RD, cough, pain, dysphagia
- Infection

Investigations CXR & CT

Treatment Surgical excision

Pulmonary AV Fistula

Definition

- The usual communication is between the pulmonary artery and pulmonary vein
- **Osler-Weber-Rendu syndrome** (hereditary hemorrhagic telangiectasia): The commonest
 - AD disease
 - Telangiectasia (Skin, GIT, nose): Epistaxis, GIT bleeding, anemia
 - AVM: Lung, liver (HF, portal HTN, encephalopathy), brain (Headache & bleeding)

Clinical Picture

- Large fistulas: RD, cyanosis, clubbing, continuous murmur, polycythemia, hemoptysis
- CNS complications: Cerebral thrombosis, abscess
- Cardiomegaly and heart failure are not present, why? "*Low pressure fistula*"

Investigation

- CXR & fluoroscopy
- Pulmonary angiography

Treatment

- Surgical excision (lobectomy)

Pulmonary Edema

Definition

- Excessive accumulation of fluid in the interstitial & intra-alveolar spaces of the lungs
- Sequences: Desaturation, ↓↓ lung compliance, RD
- Traditionally classified into: Cardiogenic & non-cardiogenic

Etiology

A) ↑↑ Pulmonary capillary pressure

- **Cardiogenic:** LV failure & mitral valve disease
- **Non-cardiogenic:** Pulmonary venoocclusive disease, mediastinal tumor

B) ↑↑ Capillary permeability (*Direct or Mediators; TNF, Histamine...*)

- Pneumonia (*Infectious*)
- ARDS
- Sepsis
- Inhaled toxic agents
- Smoke inhalation
- Aspiration pneumonia
- Radiation pneumonia
- Drowning & near drowning
- Transfusion reaction
- Uremia

C) Lymphatic insufficiency: Congenital & acquired

D) ↓↓ Oncotic pressure: Hypoalbuminemia

E) ↑↑ Negative interstitial pressure

- Upper airway obstruction
- Lung re-expansion

F) Miscellaneous causes

- Neurogenic pulmonary edema
- High-altitude pulmonary edema
- Heroin pulmonary edema
- Pancreatitis
- Pulmonary embolism
- Eclampsia

Clinical Picture (Clinical diagnosis)

- Variable degrees of **RD**
- Frothy sputum (Blood-tinged)
- Chest examination: Coarse crepitations (Bubbling) ± Wheezes

Investigations

- **CXR:** Haziness of both lung fields "butterfly pattern" (sparing the lung apices)
- **Brain natriuretic peptide:** ↑↑ in heart disease

Treatment

- ICU
- Oxygen therapy
- Diuretics: Furosemide
- Aminophylline
- Morphine (Sedation & VD)
- Positive Inotropes...
- Mask or nasal CPAP
- Mechanical ventilation: ↑↑ PEEP
- Rx of the cause
- Dialysis (Non-renal...???)

	Cardiogenic	Non-Cardiogenic
Heart size	↑↑	Normal
Vascular pedicle	↑↑	Normal
Edema	Central	Patchy
Effusion	Present	No
Septal lines	Present	No
Air bronchogram	No	Present

Community-Acquired Pneumonia

Definition

- Inflammation of the lung parenchyma
- Recurrent pneumonia: ≥ 2 attacks in 1 year or ≥ 3 attacks ever

Epidemiology

- 1st or 2nd cause of death in developing countries (19% of all < 5 yrs deaths)
- Incidence is 10 fold higher than developed countries
- Recently, there is $\downarrow\downarrow$ in pneumonia mortality (Antibiotics, vaccines)
- There is $\downarrow\downarrow$ in pneumonia caused by H.influenza, pneumococci & measles

Pathology

- Alveoli: Consolidation
- Interstitium: Infiltration with inflammatory cells

Classification

- A) **Anatomical:** Lobar, bronchopneumonia or interstitial pneumonia
 B) **Etiological:** Infectious or noninfectious ...

Etiology

- A) **Bacterial:**
- Gram +Ve: Streptococcus pneumoniae, Staphylococci, Streptococci
 - Gram -Ve: Hemophilus influenza, Klebsiella, Pseudomonas
 - TB, Mycoplasma
- B) **Viral:**
- Common:** RSV, adenoviruses, parainfluenza, influenza
 - Uncommon:** Rhinovirus, enterovirus, HSV, CMV, measles, varicella
- C) **Fungal:** Histoplasma, Aspergillus, Cryptococcus, Pneumocystis jirovecii
 D) **Parasitic:** Ascaris
 E) **Rickettsial**
 F) **Other causes (Non-infectious):**
- Aspiration pneumonia: Amniotic fluid, food, foreign body, hydrocarbons
 - Hypostatic pneumonia: Lying in one position for long time (lung congestion & edema)
 - Hypersensitivity
 - Radiation

Bacterial			
Common	Comment	Uncommon	Comment
Strept. pneumoniae	Consolidation, empyema	H. influenzae type b	Unimmunized
Group B streptococci	Neonates	Staph. aureus	Infants, pneumatoceles
Group A streptococci	Empyema	Moraxella catarrhalis	
Mycoplasma pneumonia*	Adolescents	Neisseria meningitidis	
Chlamydia pneumonia*	Adolescents	Francisella tularensis	
Chlamydia trachomatis	Infants	Nocardia species	
Mixed anaerobes	Aspiration pneumonia	Chlamydia psittaci*	
Gram-negative enterics		Yersinia pestis	Plague
		Legionella*	Contaminated water
		Coxiella burnetii*	Q fever

Streptococcus pneumoniae is the commonest bacteria in children 3 wks to 4 yrs

Mycoplasma & Chlamydia are the commonest > 5 yrs

S. pneumonia, H. influenza & S. aureus are the major causes of hospitalization & mortality

*Atypical pneumonia: may have extrapulmonary manifestations, low-grade fever, -Ve sputum & ?Rx

Clinical Picture

- History of preceding URT infection
- **Fever:** higher in bacterial causes (& chills)
- **Respiratory distress:** Tachypnea, retraction, grunting, cyanosis
- **Chest Examination:** ↓↓ Air entry, crepitations, wheezes
 - Inspection: RD
 - Palpation: TVF, position of the trachea
 - Percussion: Dullness with consolidation, collapse or effusion
 - Auscultation: ↓↓ Air entry, crepitations, wheezes

Investigations

NB: Repeat CXR is not indicated to proof cure

- CBC, ESR, CRP
- Sputum culture
- Blood culture: positive in 10% of patients with pneumococcal pneumonia
- Nasopharyngeal aspirate: for viral agents (RSV)
- Pleural fluid examination
- Serology: paired samples with 14 days interval
- CXR
 - Bacterial: Lobar consolidation (air bronchogram)
 - Staph: Pneumatocele, abscess
 - Viral: Parahilar shadows with radiating streaks
 - Mycoplasma: Patchy segmental consolidation & hilar lymphadenopathy

Causative organism

		Bacterial pneumonia	Viral pneumonia
Fever		High	Mild-moderate
Severity		More severe	Less severe
Lab.	CBC	Leukocytosis (PNLs)	Leukocytosis (Lymphocytes)
	CRP	Positive	Negative
	ESR	↑↑	Normal or mildly ↑↑

Pathological types

	Lobar pneumonia	Bronchopneumonia	Interstitial pneumonia
Site	Unilateral affection of one or more lobes	Bilateral involvement of both lungs	Bilateral involvement of interstitial tissues
Etiology	Bacterial Mycoplasma	Bacterial Viral	Mostly viral
Chest Ex.	Auscultation: ▪ ↓↓ Air entry ▪ Bronchial breathing	Auscultation: ▪ Fine consonating crepitations	Auscultation: ▪ Expiratory wheezes
CXR	Lobar consolidation	Fine nodular or Patchy infiltration	Dense parahilar shadows with radiating streaks

Treatment

Indications of Hospitalization in children with pneumonia

1. Age < 6 months
2. Immunocompromised: 1ry, 2ry, sickle cell disease
3. Toxic appearance
4. Severe RD
5. Vomiting
6. Dehydration
7. Requirement for supplemental O₂ therapy (O₂ Saturation < 90% in room air)
8. Social factors: Non-compliance...

A) Mild pneumonia in older children: Home management

1. Close observation (Degree of RD)
2. Antibiotics

2mo-5y	≥ 5y
-Oral Amoxicillin 80mg/kg/day in 3 divided doses for at least 7 days -Alternatives ➤ Amoxicillin/Clavulinate ➤ Oral cephalosporins ➤ Macrolides -Consider 24 hours of a parenteral 3rd generation cephalosporin in patients with more severe disease: ➤ Ceftriaxone 50 mg/Kg once, or ➤ Cefotaxime 50-100 mg/Kg/day	Macrolides: Erythromycin 40 mg/kg/day for ≥ 7 days or Azithromycin 10 mg/kg/day for 3-5 days

B) Moderate to Severe pneumonia: Hospital management

a. Non specific support

1. Oxygen therapy
2. Suction of airway secretions
3. Physiotherapy
4. Proper hydration: IV fluid
5. Mechanical ventilation may be needed in severe cases

b. Specific

1. Parental antibiotic therapy
2. Antibiotics should be modified according to the results of culture & sensitivity

Neonates	1mo- 5 yrs	≥ 5y
I.V. Ampicillin (100 mg/Kg/day) +Gentamycin (5 mg/Kg/day)	Parenteral 3 rd generation cephalosporin	I.V. Ampicillin or Parenteral 3 rd generation cephalosporin + Macrolide
Critically ill		
Add 3 rd generation cephalosporin	Parenteral cephalosporin + Amoxicillin/ Clavulinate	Parenteral 3 rd generation cephalosporin + Macrolide

Complications

1. Respiratory failure: Most serious
2. Pleural effusion: Bacteria specially Staphylococci & Streptococcus pneumoniae
3. Lung abscess: Bacteria specially Staphylococci
4. Pneumothorax: Bacteria specially Staphylococci & Klebsiella
5. Myocarditis & Acute HF: In infants with severe bacterial infection
6. Hematologic spread: Meningitis, arthritis, osteomyelitis...
7. Complications of mycoplasma
 - **Bacterial superinfection**
 - **CNS:**
 - Meningoencephalitis
 - Aseptic meningitis
 - GBS & transverse myelitis
 - Ataxia
 - **Hematological:**
 - AIHA (Cold hemagglutinins)
 - Thrombocytopenia
 - **Skin:**
 - Erythema multiforme
 - Stevens-Johnson syndrome
 - **Others:**
 - Hepatitis
 - Pancreatitis
 - Arthritis
 - Myocarditis & Pericarditis

Prevention

Important Remarks about some bacterial pneumonia

Organism	Comment
Hemophilus influenza pneumonia	Usually lobar Complications: meningitis (CSF?) Rx: Cefotaxime or ceftriaxone
Staphylococcal pneumonia	Usually bronchopneumonia Unilateral or more prominent on one side Complications: Cavitation, abscess formation, pneumatocele, pneumothorax, pleural effusion & empyema Rx: Amoxicillin-clavulanic acid Ampicillin-sulbactam Oxacillin Vancomycin or clindamycin
Pneumococcal pneumonia	Usually bronchopneumonia Complications: Pleural effusion & empyema Rx: Penicillin G
Klebsiella pneumonia	Usually bronchopneumonia Complications: As staphylococcal pneumonia

Slowly resolving pneumonia

Definition

- Persistence of symptoms or radiographic findings beyond the expected time course
- Typically, improvement of clinical symptoms occurs within 48-96 hrs after Rx

Etiology

1. Complications: empyema
2. Bacterial resistance
3. Non-bacterial causes: TB & viruses
4. Noninfectious causes: Bronchiolitis obliterans, Wegener granulomatosis
5. Bronchial obstruction: Foreign body, mucous plugs...
6. Immunodeficiency
7. Lung disease: Immotile cilia syndrome, cystic fibrosis

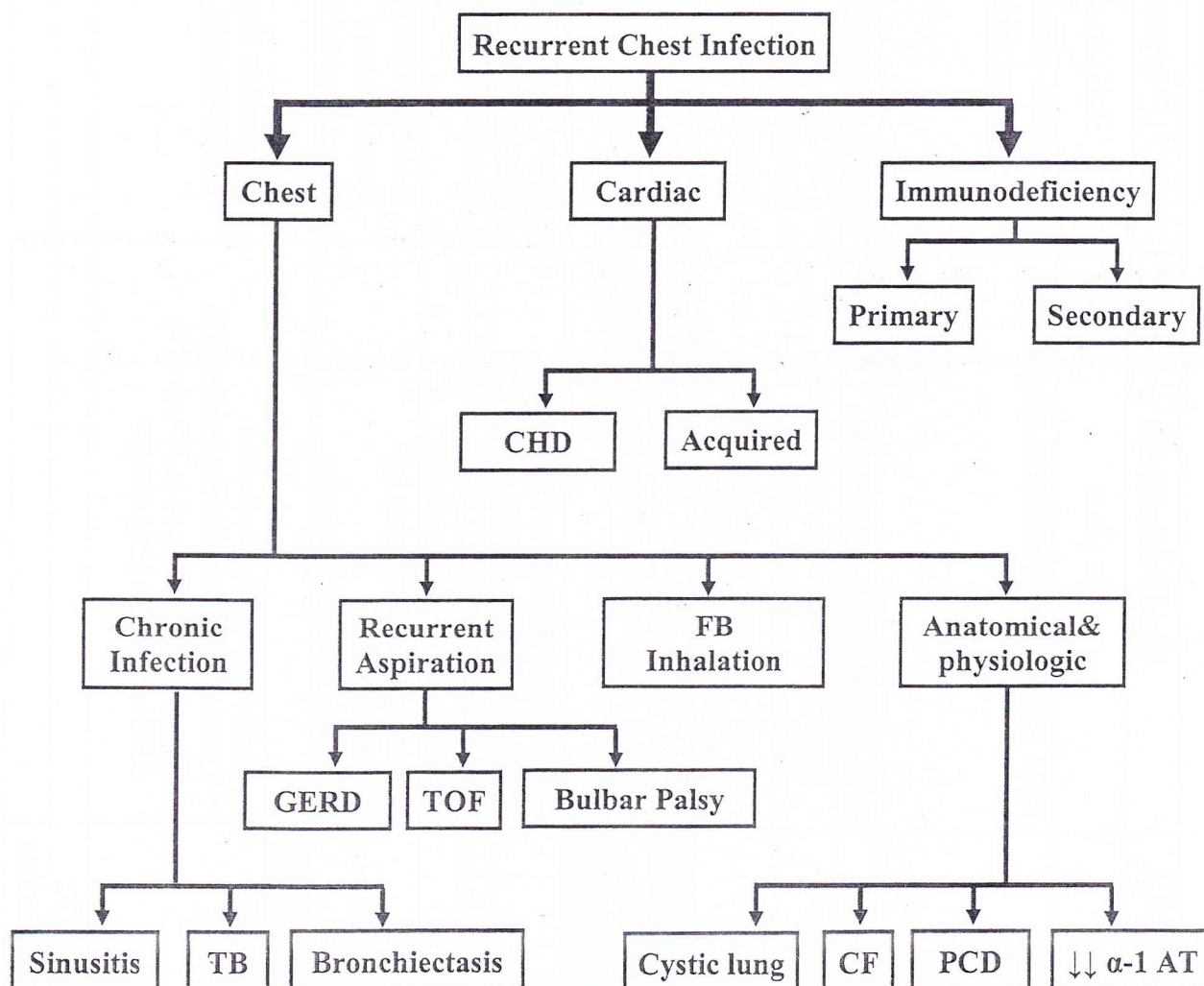
Evaluation

- Repeat the CXR is the 1st step
- Investigations of the possible etiology: CT chest, bronchoscopy, ANCA...

Recurrent Chest Infection

Normal RCI Up to 6-8 acute respiratory illnesses per year is not unusual?!

Abnormally RCI



Pneumonia

	Viral Pneumonia	Mycoplasma	Bacterial Pneumonia
Age	2-3 years	School-aged children	- Infancy: Staphylococci - 2-5 yrs: Streptococci (Group A)
Etiology	RSV, Parainfluenza, influenza, adenoviruses	Mycoplasma pneumoniae "The <u>smallest self-replicating agents</u> "	- Streptococcus pneumoniae - Staphylococci - Streptococci - Hemophilus influenza type b - Chlamydia
Pathology	Bronchopneumonia or interstitial	Bronchopneumonia or interstitial	Lobar or Bronchopneumonia
Symptoms	▪ Preceding upper respiratory tract symptoms; cough & rhinitis ▪ Fever is < bacterial pneumonia ▪ Variable degrees of RD	▪ FAHM ▪ Cough usually ↑↑ during the 1st week ▪ Symptoms severity > Physical signs	▪ Preceding upper respiratory tract symptoms ▪ Fever: High ± chills ▪ Variable degrees of RD
Chest signs			
Auscultation	Normal or scattered crepitations & wheezes	Crepitations	Localized bronchial breathing & crepitations
Investigations			
CXR	- Bronchopneumonia or interstitial - Hilar lymphadenopathy	- Patchy segmental consolidation - Hilar lymphadenopathy	- Lobar consolidation - Bronchopneumonia
CBC, CRP, ESR	- Normal or mildly ↑↑ TLC - Lymphocyte predominance	- Normal TLC	- Leukocytosis (↑↑ TLC) - PNLS predominance
Other Investigations	- Serology: IgM or rising IgG - Viral culture - Detection of viral antigens	- Serology: IgM or rising IgG - Sputum culture - Direct coombs' test & reticulocytosis	- Sputum: Culture & Sensitivity - Blood: Culture & Sensitivity - Pleural tap: Cytology & culture
Treatment			
Medical Rx	▪ Antibiotics are not indicated ▪ Coexisting bacterial infection: in 30% ▪ Ribavirin may be used in infants with CHD or chronic lung disease	▪ Macrolides: Erythromycin, Clarithromycin, Azithromycin	▪ Mild cases: Amoxicillin (± clavulanic acid) ▪ Severe cases: Parenteral Antibiotics; cephalosporins (Cefotaxime, ceftriaxone) ▪ Staphylococci: Vancomycin or clindamycin ▪ Modify antibiotics according to C & S
Surgical Rx			▪ Intercostal tube drainage under water seal
Complications			

Chronic Recurrent Aspiration

Definition

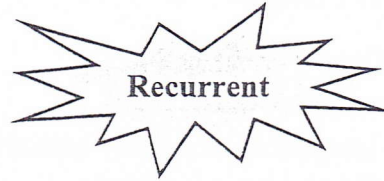
- Recurrent aspiration of gastric, nasal, oral secretion
- Wide clinical spectrum; asymptomatic to life-threatening events

Pathology

- Granulomatous inflammation
- Interstitial inflammation & fibrosis

Effects of Aspiration

- Cough, bronchitis & bronchiolitis
- Pneumonia, wheezes, atelectasis, laryngospasm



Predisposing Factors of Aspiration

A) Anatomical

- | | |
|---|---|
| ☒ Micrognathia & Macroglossia | ☒ N/G tube, ETT, tracheostomy |
| ☒ Cleft palate | ☒ Achalasia |
| ☒ Tracheoesophageal fistula | ☒ Scleroderma |
| ☒ Vascular ring | ☒ GERD & hiatus hernia |

B) Neuromuscular (Esophageal dysmotility)

- | | |
|---|--|
| ☒ Cerebral palsy & Chiari malformation | ☒ Myasthenia gravis |
| ☒ Stroke | ☒ Muscular dystrophies |
| ☒ Multiple sclerosis | ☒ Polio, SMA |
| ☒ Cricopharyngeal achalasia: Failure of relaxation of UES | |
| ☒ Cricopharyngeal incoordination: Incoordination of contraction | |

C) Miscellaneous

- ☒ Poor oral hygiene
- ☒ Poor feeding techniques: overfeeding, inappropriate foods...

Approach to a child with frequent aspiration

Investigations

- CXR: Infiltration, atelectasis
- HRCT chest
- Barium swallow: Useful in...
- Videofluoroscopic swallow study (VSS): Gold standard for evaluation of swallowing
- Esophageal pH monitoring
- MII
- Gastroesophageal scintigraphy
- Radionuclide salivagram
- Fiberoptic endoscopic evaluation of swallowing (FEES)
- Bronchoscopy (BAL): Lipid-laden macrophages

Treatment

- Treatment of the cause
- Supportive Rx: Proper position, thickened foods, ↓↓ amount/feed, NG tube, postpyloric
- Medical Rx: Anticholinergic
- Surgical Rx: Fundoplication with gastrostomy tube, salivary gland excision

Pulmonary Hemosiderosis

(Diffuse alveolar hemorrhage = DAH)

Definition

- Triad of iron-deficiency anemia, hemoptysis & lung infiltrates (CXR)

Epidemiology

- IPH: 1: 1 million

Etiology

A) Primary hemosiderosis

- Idiopathic pulmonary hemosiderosis (IPH)
- Goodpasture syndrome
- Heiner syndrome (Cow's milk allergy)

B) Secondary hemosiderosis

- Congestive HF, mitral stenosis
- Pulmonary HTN, AVM, pulmonary VOD, pulmonary lymphangioleiomyomatosis
- Collagen-vascular diseases: SLE, HSP, APS, JRA, PAN, Wegener granulomatosis, microscopic polyangiitis
- Coagulopathy
- HUS, CRF, IgA nephropathy
- Immunodeficiency: CGD
- Drug-induced capillaritis
- Prematurity: immature alveoli & vessels
- Child abuse

Pulmonary hemosideosis:

a. With pulmonary capillaritis

- Collagen-vascular
- Goodpasture syndrome
- Drug-induced capillaritis

b. Without pulmonary capillaritis

- Cardiac: HF, MS, pulmonary HTN
- Noncardiac: IPH, Heiner, ID

Pathology

- Repeated episodes of pulmonary Hemorrhage
- Hemosiderin deposition $\Rightarrow$ Hemosiderin-lade macrophages (HLMs)
- Hemosiderin is formed within 72 hrs & HLMs last for weeks after hemorrhagic event
- Inflammation (mediators) & neutrophil influx
- Thickening of alveolar septa & finally, pulmonary fibrosis

Clinical Picture (Variable)

- Variable: Asymptomatic $\longleftrightarrow$ Shock & sudden death
- Hemoptysis may not occur!!
- RD, cough, wheezes, why?
- General examination: Pallor, tachycardia, RD, fever, shock (severe hemoptysis)

Investigations

- **CBC:** Microcytic hypochromic anemia, eosinophilia
- **Retics & Bilirubin:** $\uparrow\uparrow$
- **Iron studies:** $\downarrow\downarrow$ iron, $\uparrow\uparrow$ TIBC
- **ANA, Anti DNA, APA, ANCA, Anti-GBM**
- **Stools:** Occult blood in stools
- **Sputum & BAL:** HLMs
- **CXR:** lung infiltrates "butterfly pattern" (sparing the lung apices)
- **CT chest**
- **Pulmonary function tests:** Obstructive changes (Late: restrictive)
- **DLCO:** $\uparrow\uparrow$, why?

Treatment

- **Supportive therapy:** Oxygen therapy, ventilator support, IVF, blood transfusion
- **IPH:** Steroids (2 mg/Kg/d) ± immunosuppressives
- **Lung transplantation**

Pulmonary Hge & Hemoptysis

Definition

- Pulmonary hemorrhage; hemoptysis may be absent

Epidemiology Underestimated, why?

Etiology

A) Focal hemorrhage

- Infection: Bronchitis, bronchiectasis, CF
- TB
- Trauma: tracheostomy
- Tumors (Hemangioma)
- Pulmonary AV malformation
- Pulmonary embolism
- FB

Focal hemorrhage can be treated by embolization or lobectomy

B) DAH: See before

C) Acute idiopathic pulmonary hemorrhage (AIPH): In infants ≤ 1 yr

Atelectasis

Definition

- Incomplete expansion or collapse of lung tissue with decreased aeration
- It may be segmental, lobar or complete lung collapse
- 90% affecting the upper lobes (Rt > Lt)

Etiology

Causes		Examples
External compression		Pleural effusion, pneumothorax, diaphragmatic hernia
Obstruction	Obstruction (Lumen)	FB, granulomatous tissue, mucous plugs, broncholithiasis, ETT
	Obstruction (Wall)	LN, stricture, bronchiolitis, asthma
	Obstruction (Outside)	LN, tumor, cardiomegaly, pulmonary abscess, vascular ring
Respiratory paralysis		Neuromuscular abnormalities, restrictive casts & dressings, osseous deformities, defective movement of the diaphragm

Clinical Picture

- Variable degrees of RD (asymptomatic if small)
- Chest examination: (Inspection...) Retraction, mediastinal shift, Dullness, ↓↓ air entry

Investigations CXR, CT, Bronchoscopy

Treatment Respiratory support, physiotherapy, suction, DNase, CPAP

Pneumothorax

Definition

- It is the presence of air within the pleural space

Incidence

- Asymptomatic pneumothorax affects up to 1-2% of all newborn infants

Etiology

A) Spontaneous

a. Primary:

- No trauma, No lung disease
- Reported in some families (Folliculin gene)

b. Secondary

- ☒ **Congenital lung diseases:** Lung hypoplasia, CCAM, CLE, bronchogenic cyst
- ☒ **Infections:** Pneumatocele, lung abscess
- ☒ **Conditions with ↑↑ intrathoracic pressure:** Asthma, bronchiolitis, CF, FB, MAS
- ☒ **Diffuse lung diseases:** Ehlers-Danlos, Marfan, Langerhans cell histiocytosis
- ☒ **Metastatic:** Osteosarcoma*

B) Traumatic

a. Non-iatrogenic: Blunt or penetrating injury

b. Iatrogenic

- **Barotrauma** (ambu-bag or mechanical ventilation)
- Vigorous resuscitation or physiotherapy
- Vascular access insertion "pleural injury"
- Chest surgery, tracheostomy, thoracocentesis

Barotrauma:

1. ↑↑ Pressure (PIP or PEEP)
2. ↑↑ IT
3. ↓↓ Expiratory time
4. ETT
5. Improved pathology without...
6. Unilateral lung pathology
7. Inadequate humidification
8. Aggressive physiotherapy

Pathogenesis

- Hypoxemia (V/Q mismatch)
- Simple pneumothorax: Intrapleural pressure is atmospheric
- Tension pneumothorax: IPP is ↑↑ (Compression, shift, ↓↓ VR & CO, Obstructive shock)

Clinical Picture

- Variable degrees of RD (asymptomatic if small)
- Chest examination: Inspection...
- Chest examination: Unilateral bulge, mediastinal shift, hyper-resonance, ↓↓ air entry
- May be bilateral in 10%
- Tension pneumothorax (↓↓ VR, ↓↓ BP): Obstructive shock

Investigations

- CXR: Jet-black translucency + Mediastinal shift (may be absent, when?)
- Needle aspiration (2nd intercostals space MCL): "Diagnostic Therapeutic test"

Treatment

- Conservative management (if asymptomatic): Observation + 100 % O₂
- Needle aspiration
- Chest tube with underwater-seal drainage
- HFV is the ventilatory treatment of choice

Complications

- Respiratory failure
- Obstructive shock
- SIADH
- ICH (↑↑ Intra-thoracic pressure)

Pulmonary Embolism & Infarction

Incidence

- 1:100.000

Etiology (Risk Factors)

1. **Environmental** Smoking, immotility
2. **Women's health:** OCPs, Pregnancy, hormonal replacement therapy
3. **Surgical**
 - Trauma
 - Orthopedic, neurosurgical & general surgery
4. **Thrombophilia:**
5. **Medical conditions**
 - DM, HTN, HF, obesity
 - Central venous catheter
 - Malignancy
 - Permanent pacemaker
 - Intracardiac defibrillator
6. **Non-thrombotic**
 - Air, Amniotic fluid, fat, bone, tumors, FB

Pathogenesis

- V/Q mismatch (Dead space) & hypoxia
- Hyperventilation & hypocapnia
- Pulmonary infarction is unusual, why?
- Pulmonary HTN & cor pulmonale

Clinical Picture

- Picture of the cause
- Variable presentation (may be asymptomatic)
- Pain, dyspnea, cough
- RVF
- Chest examination: localized crackles may be detected

Investigations

- CXR: Non-specific
- Spiral CT angiography
- Pulmonary angiography
- EEG: RV strain
- Echo-:
- ABG: Hypoxia
- Lung scan
- Investigations of the cause

Treatment

- Stabilization
- Anticoagulation: Heparin or LMW heparin followed by oral anticoagulant
- Thrombolytic therapy
- Surgical embolectomy

Prognosis

- Mortality: 2.2%

Parenchymal Diseases with prominent Hypersensitivity, Eosinophilic infiltration

Hypersensitivity pneumonitis

(Extrinsic allergic alveolitis)

Definition

- Immune-mediated diffuse inflammatory disease of the pulmonary interstitium caused by inhalation of organic antigens (typically animal or vegetable origin but may be drug-related)

Etiology & Types of HP

- **Farmer's lung**: From moldy hay
- **Bird fancier "Breeder lung"**: From the feces or urine of some bird & animal species
- **Ventilation "humidifier HP"**: From contaminated humidifiers or air conditioners
- **Pigeon breeder disease**
- **Bagassosis**: Sugarcane
- **NB**: The usual antigen is thermophilic actinomycetes
- **NB**: Feather filled bedding (pillows, quilts) can cause the disease

Pathogenesis

- **Acute**: Infiltration by PNLs, lymphocytes, plasma cells & macrophages
- **Subacute**: Noncaseating granuloma & bronchiolitis
- **Chronic**: Noncaseating granuloma, interstitial fibrosis & honeycombing
- **Mechanism**: Type III & IV hypersensitivity reactions

Clinical Picture (High index of suspicion)

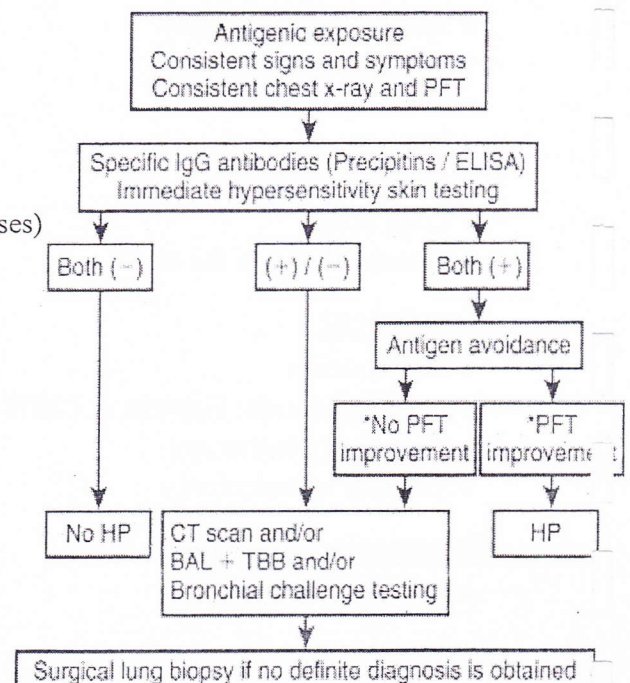
- **Acute** (*within 4-8 hr after exposure*): Fever, chills, cough, dyspnea, myalgia & malaise
- **Subacute**: Cough, dyspnea, anorexia & weight loss
- **Chronic**: Cough, dyspnea, anorexia, weight loss, hypoxemia, clubbing & cor pulmonale
- **Major criteria**: Compatible C/P, exposure (antibody studies), compatible CXR or HRCT, BAL (lymphocytes), compatible histology & positive antigen provocation challenge
- **Minor criteria**: Bibasilar crackles, ↓↓ diffusion capacity & hypoxemia

Investigations

- CBC: leukocytosis, neutrophilia, shift to left
- Serum IgG, IgM & IgA: ↑↑
- Specific IgG antibody studies
- Skin testing to particular antigens
- Inhalational challenge: certain centers
- CXR: ground-glass haziness (sparing lung apices & bases) reticulonodular
- HRCT: may show bronchiectasis
- PFT: Restrictive pattern

Treatment

- Avoidance
- Prednisone (1-2 mg/kg/24 hr) for 6-12 mo



Silo Filler Disease

(Silage gas poisoning)

Definition

- Lung disease caused by nitrogen dioxide toxicity which is produced in silos
- Dangerous concentrations of gas can remain in a closed silo for $\approx$ 2 wk

Pathogenesis

- Inhalation, chemical pneumonitis, airway epithelial hyperplasia & pulmonary edema
- Methemoglobinemia in severe cases

Clinical Picture

- Cough, dyspnea, wheezes
- Variable: Fatal, complete recovery, chronic lung disease (fibrosis & emphysema)

Investigations

- CXR: Pulmonary edema

Treatment

- High-dose prednisone
- Methemoglobinemia: Methylene blue

Paraquate Lung

Definition

- Paraquat is the most toxic dipyridilium herbicide

Pathogenesis

- Production of superoxides & free radicals leading to mitochondrial damage & cell death
- Paraquat-induced injury is increased in the presence of high O_2 concentrations

Clinical Picture (Dose-related)

- Asymptomatic
- Vomiting, diarrhea & caustic injury to the oral or esophageal mucosa
- Organ failure: Renal failure, hepatic dysfunction, pulmonary fibrosis, respiratory failure

Investigations

- Plasma paraquat concentration (paraquat nomograms): Diagnostic & prognostic
- Urine dithionite test (turns urine blue if paraquat is present)

Treatment

- Supportive
- Antioxidant therapy: Deferoxamine & acetylcysteine
- Immunosuppressive therapy consisting of IV steroids & cyclophosphamide
- Lung transplantation: Poor outcome, why?

Prognosis

- Poor

Eosinophilic lung Disease

(Pulmonary infiltrates with eosinophilia (PIE) or Löffler Syndrome)

Definition

- Heterogenous group of disorders characterized by pulmonary infiltrates & circulating or tissue eosinophilia

Epidemiology

- Rare in children (Only 6% of cases occurred in patients <20 yr of age)

Etiology

A) Primary (Idiopathic)

- ☒ Simple pulmonary eosinophilia (Löffler syndrome)
- ☒ Acute eosinophilic pneumonia
- ☒ Chronic eosinophilic pneumonia
- ☒ Idiopathic hypereosinophilic syndrome

B) Secondary

- ☒ Tropical pulmonary eosinophilia
- ☒ Pulmonary eosinophilia with asthma
- ☒ Polyarteritis nodosa
- ☒ Churg-Strauss syndrome
- ☒ Allergic bronchopulmonary aspergillosis (ABPA)
- ☒ Drug-induced eosinophilic lung disease

Pathogenesis

- The most common **etiologies** of PIE syndromes are parasite infections and drug reactions
- **Parasites:** *Ascaris lumbricoides*, *Strongyloides*, *Toxocara canis*, *Wuchereria bancrofti*
- **Drugs:** Sulfasalazine, penicillin, ampicillin, ibuprofen & cromolyn
- **Pathology:** Alveolar macrophages, lymphocytes, neutrophils & eosinophils

Clinical Picture

- Cough, dyspnea, wheezes, malaise, fever
- In acute eosinophilic pneumonia, symptoms last for < 1 month
- In chronic eosinophilic pneumonia, symptoms last for > 6 months

Investigations

- CBC: Eosinophilia
- CXR: Bilateral & diffuse nonspecific interstitial, alveolar, or mixed infiltrates
[NB: Chronic eosinophilic pneumonia: photographic negative of pulmonary edema]
- BAL: Eosinophilia
- Lung biopsy

Treatment

- Avoidance of exposure
- Rx of parasitic infestations
- Acute & chronic eosinophilic pneumonia: prednisone

Prognosis

- Generally good
- Acute eosinophilic pneumonia may be fatal

Interstitial Lung Diseases

Definition

- Heterogenous group of disorders affecting the interstitium (the tissue around the alveoli)
- It involves alveolar epithelium, pulmonary capillary endothelium & basement membrane
- Some cases are familial (surfactant dysfunction disorders)

Epidemiology

- Rare in children

Etiology

A) Disorders more common in infants

- ☒ Developmental disorders: acinar dysplasia, congenital alveolar dysplasia, alveolar capillary dysplasia
- ☒ Growth disorders (deficient alveolarization): Pulmonary hypoplasia, CHD, chromosomal disorders
- ☒ Surfactant dysfunction: SP-B mutation, SP-C mutation, ABCA3 mutation
- ☒ Pulmonary interstitial glycogenosis
- ☒ Neuroendocrine cell hyperplasia of infancy

B) Disorders of known association

- ☒ Hypersensitivity pneumonitis
- ☒ Aspiration syndromes
- ☒ Infections: adenoviruses, influenza viruses, Chlamydia, Mycoplasma

C) Disorders of immunocompromised host

- ☒ Opportunistic infections
- ☒ Therapeutic interventions: chemotherapy, radiation, transplantation

D) ILD of unknown etiology

- ☒ Usual interstitial pneumonitis (UIP)
- ☒ Desquamative interstitial pneumonitis (DIP)
- ☒ Lymphocytic interstitial pneumonitis (LIP)
- ☒ Nonspecific interstitial pneumonitis (cellular/fibrotic)
- ☒ Eosinophilic pneumonia
- ☒ Pulmonary hemosiderosis
- ☒ Pulmonary alveolar proteinosis
- ☒ Pulmonary vascular disorders
- ☒ Pulmonary lymphatic disorders
- ☒ Pulmonary microlithiasis

E) Systemic disorders with pulmonary involvement

- ☒ Collagen-vascular disorders
- ☒ Malignancies
- ☒ Langerhans cell histiocytosis
- ☒ Sarcoidosis
- ☒ Neurocutaneous syndromes: NF & TS
- ☒ Storage diseases: Gaucher & Niemann-Pick
- ☒ Vasculitis: Wegener & Churge-Strauss syndrome

Clinical Picture

- Symptoms are usually insidious and continuous (Not episodic)
- Cough, dyspnea, FTT, clubbing, cyanosis (Wheezing and fever are **not** common)
- Hypoxemia (Hypercarbia is late, why?)
- Later: pulmonary HTN, cor pulmonale
- Picture of the cause: pulmonary hemosiderosis...

Investigations

- CXR & CT chest: interstitial, reticular, nodular, reticulonodular, or honeycombed
- HRCT chest: Better information
- PFTs: restrictive pattern
- BAL: Pulmonary alveolar proteinosis & pulmonary hemosiderosis
- Lung biopsy

Treatment

- Supportive care: O₂ therapy (hypoxia), adequate nutrition, antimicrobial Rx (infections)
- Some cases may respond to bronchodilators
- Corticosteroids: treatment of choice (*1-2 mg/kg/24 hr for 6-8 wk with gradual tapering*)
- Other lines: Azathioprine, cyclophosphamide, cyclosporine, hydroxychloroquine, MTX, IVIG, GM-CSF & pulse high-dose steroids
- Lung transplantation

Prognosis

- Some children recover spontaneously, but others progress to death
- Pulmonary HTN, FTT & severe fibrosis are poor prognostic indicators

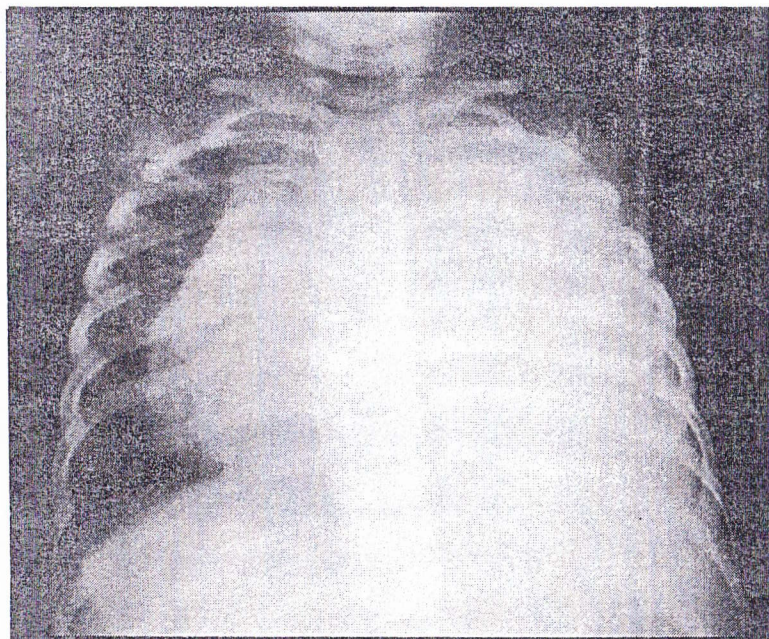
Suppurative Lung Syndromes

Definition

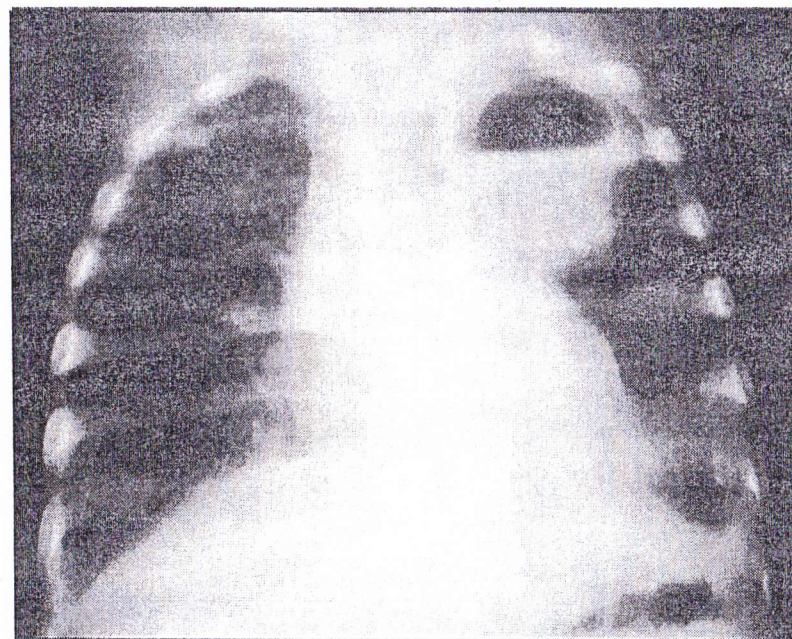
- Group of diseases characterized by suppurative inflammation of the pleura, lung or bronchi

	Empyema	Lung abscess	Bronchiectasis
Definition	Accumulation of pus in the pleural cavity	Localized suppurative inflammation resulting in a cavity containing pus	Permanent dilatation of the bronchi with suppurative inflammation of their walls
Etiology	▪ Bacterial pneumonia (Staph., Streptococcus pneumoniae, H. influenza) ▪ Trauma ▪ Mediastinitis ▪ Intra-abdominal abscess	▪ Bacterial pneumonia (Staph., Klebsiella) ▪ Aspiration of infected material ▪ FB ▪ Metastatic lung abscess (Rt sided infective endocarditis) ▪ Amebic liver abscess	A) Congenital: Abnormal bronchial development B) Acquired: Chronic pulmonary infection ▪ Immunodeficiency ▪ Cystic fibrosis ▪ Immotile cilia syndrome ▪ Asthma ▪ Aspiration: FB, TOF, GERD
Pathology	- Pleura: Inflammation - Pleural space: Pus	- Cavity containing pus - Fibrosis	- Site: Usually bilateral & basal - Types: Cylindrical, saccular or fusiform - Mucosa: Ulceration + loss of ciliated epith. - Bronchial wall: Infiltration, edema, fibrosis - Lumen: Mucus, pus ± blood
Symptoms	▪ Fever, weight loss ▪ Cough, dyspnea ▪ Lying on the affected side ▪ Pain	▪ FAHM, weight loss ▪ Cough, hemoptysis ▪ Expectoration of amount of purulent sputum followed by ▪ Marked relief of symptoms	▪ FAHM, weight loss ▪ Cough, dyspnea, expectoration of excessive amount of muco-purulent sputum (Changing with posture) ▪ Hemoptysis, RD, clubbing
Chest signs			
Inspection	Unilateral bulge ↓↓ Respiratory movement	Normal	Normal
Palpation	Trachea & heart may be shifted to the opposite side	Central mediastinum (Trachea & heart)	Central mediastinum (Trachea & heart)
Percussion	Stony dullness (on the affected side)	Localized dullness	Patchy dullness
Auscultation	↓↓ Air entry (on the affected side)	Localized bronchial breathing	Localized bronchial breathing & crepitations

Investigations			
CXR	- Homogenous opacity obliterating C/P angle & rising to the axilla - Mediastinal shift (Heart & Trachea)	- Cavity ± Air-fluid level - No mediastinal shift (DD: Hydropneum.)	- Patchy areas of consolidation - Honey-comb appearance (Dilated bronchi)
Other Investigations	CBC, ESR, CRP Thoracocentesis: Culture & Sensitivity	CBC, ESR, CRP Sputum: Culture & Sensitivity	CBC, ESR, CRP Sputum: Culture & Sensitivity HRCT, bronchography, bronchoscopy
Treatment			
Medical Rx	▪ Antibiotics: according to C & S	▪ Antibiotics: according to C & S	▪ Antibiotics: according to C & S ▪ Postural drainage & physiotherapy ▪ Supportive: Expectorants & mucolytics
Surgical Rx	▪ Intercostal tube drainage under water seal ▪ Fibrinolytic agent (<i>in the tube</i>) ▪ VATS ▪ Open decortication	▪ Bronchoscopy: Removal of FB ▪ Lobectomy: indicated if there is recurrent hemoptysis or resistant recurrent infection	▪ Lobectomy: if localized



Pleural effusion



Lt upper lobe lung abscess

Bronchiectasis

Definition

- Permanent dilatation of the bronchi with suppurative inflammation of their walls

Epidemiology

- Incidence is decreasing in developed countries
- Male : female ratio is 2 : 1

Etiology

A) Congenital:

- **Williams-Campbell syndrome:** Absence of annular bronchial cartilage
- **Marnier-Kuhn syndrome** (congenital tracheobronchomegaly): CT disorder Primary

B) Acquired: Gradually progressive

- Cystic fibrosis is the most common cause
- Primary ciliary dyskinesia
- Foreign body inhalation
- Aspiration of gastric contents
- Immune deficiency syndromes (especially humoral immunity)
- Infection: Pertussis, measles, rubella, RSV, TB
- **Rt middle lobe syndrome:** Compression of Rt middle lobe bronchus by hilar LN

Pathogenesis & Pathology

- Mechanism: Obstruction + Infection + Inflammation
- Site: Usually bilateral & basal
- Types: Cylindrical, varicose (Dilatation with local constrictions), saccular (cystic) or fusiform
- Mucosa: Ulceration + loss of ciliated epithelium
- Bronchial wall: Infiltration, edema, fibrosis
- Lumen: Mucus, pus ± blood

Clinical Picture

- FAHM, weight loss
- Cough, dyspnea, expectoration of ↑↑ amount of muco-purulent sputum (Postural)
- Hemoptysis, clubbing

Investigations

- Culture: deep throat, sputum or BAL fluid
- CXR & CT chest: Nonspecific
- Thin-section HRCT: "Gold standard" ➡
- Investigation of the cause: sweat Cl test



Treatment

- **Medical:** Chest-physiotherapy (postural drainage), antibiotics & bronchodilators
- Long-term prophylactic: Oral (macrolide) or nebulized antibiotics
- **Surgical (Lobectomy):** Localized resistant bronchiectasis
- Lung transplantation

Pulmonary Abscess

Definition

- Localized suppurative inflammation (Cavity containing pus)

Classification

- **Primary lung abscess:** occurs in a previously healthy patient (Rt*)
- **Secondary lung abscess:** with underlying or predisposing conditions (Lt*)

Etiology

A) Organisms:

- Anaerobic bacteria: Bacteroides, Fusobacterium, Peptostreptococcus
- Aerobic bacteria: Streptococcus, Staph. aureus, E. coli, Klebsiella, Pseudomonas
- Combined viral-bacterial infection
- Fungi: in immunocompromised patient

B) Predisposing factors:

- Aspiration: GERD, TOF, neurologic diseases, seizures
- Foreign body inhalation
- Postoperative complications of adeno-tonsillectomy
- Pneumonia
- Cystic fibrosis
- Immunodeficiency

Clinical Picture

- FAHM, weight loss
- Cough, dyspnea, hemoptysis
- Expectoration of ↑↑ amount of -purulent sputum followed by marked relief of symptoms

Investigations

- Culture: Sputum (usually contaminated), BAL fluid, direct lung puncture, percutaneous or transtracheal aspiration
- CXR & CT: Thick-walled cavity with air-fluid level (DD: Pneumatocele; thin-walled)

Treatment (Conservative management is recommended)

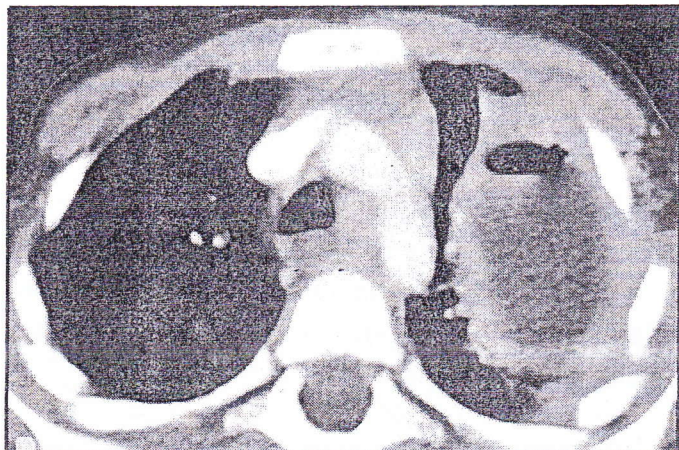
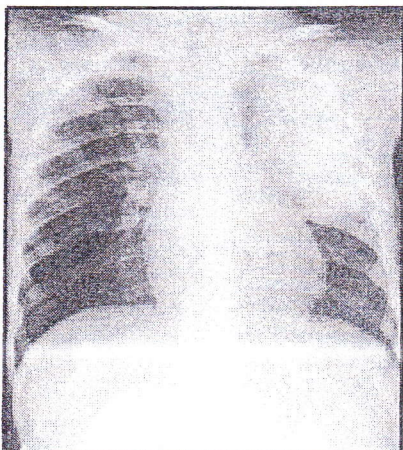
• Antibiotics (4-6 wks):

- Parenteral for 2-3 wk, followed by oral antibiotics
- Choice: Anti-Staph & anaerobic (Clindamycin)

Empiric therapy can be initiated in the absence of culturable material

• Surgical:

- Indication: failure to improve after 7-10 days of appropriate antimicrobial therapy
- Options: CT guided percutaneous aspiration, thoracotomy, lobectomy, decortication



Dry Pleurisy

Definition Inflammation of the pleura

Etiology

1. Infections

- **Viral:** Coxsackievirus B, Adenovirus
- **Bacterial:** Streptococci, Staphylococci, Pneumococci, Meningococci & Mycoplasma
- TB

2. Collagen-Vascular

- JRA
- Scleroderma
- Vasculitis Syndromes
- SLE
- Rheumatic fever
- Sarcoidosis

3. Familial Mediterranean Fever

4. Traumatic

5. Tumors: Primary or metastases

6. Post-pericariotomy syndrome

7. Metabolic: Uremia, hypothyroidism

Clinical Picture

- Pain: Stitching, localized ↑↑ with breathing & coughing and referred to the shoulder
- Position: lying on the affected side
- Pleural rub
- Other manifestations: FAHM, cough, dyspnea

Investigations CXR: Normal or may show small amount of pleural effusion

Treatment Analgesics: NSAIDs

Pleural Effusion

Definition Accumulation of fluid in the pleural space

Etiology

A) Pleural exudate: *The same causes...*

B) Hydrothorax (Transudate)

- Cardiac (HF), Renal (NS), Hepatic (LCF), Nutritional, Hypoalbuminemia
- Vascular obstruction (SVC)

C) Hemothorax

- Trauma
- VA insertion
- Tumors
- Pulmonary AV fistula
- TB
- Bleeding tendency

D) Chylothorax

E) Empyema

Clinical Picture

- Pain: Dull
- Position: lying on the affected side
- Other manifestations: FAHM, cough, dyspnea
- Examination: Bulge, shift, dullness, ↓↓ air entry

Investigations

- CXR & US: Pleural effusion...
- **Thoacocentesis:** Obtaining fluid from the pleural cavity
Needle puncture should be along the upper border of the rib, why?
Complications: Infection, bleeding, pneumothorax, liver/spleen injury

Pleural fluid examination (Physical, Chemical, Cytological & Bacteriological)

- Physical: Clear in transudate, turbid in others
- Cytological: ↑↑ PNLs (Exudate), ↑↑ Lymphocytes (TB), Malignant cells
- Chemical: Milky fluid with ↑↑ TAG (Chylous), blood-stained (Tumors)
- Bacteriological: Culture

	Transudate	Exudate
Aspect	Clear	Turbid
Specific gravity	Low (< 1015)	High (> 1015)
Proteins	Low (< 2.5 g/dl)	High (> 2.5 g/dl)
Cell Number	Few (< 2000 /mm ³)	Many (> 2000 /mm ³)
Cell Type	Lymphocytes	PNLs
LDH	Low (< 200 IU/L)	High (> 200 IU/L)
pH	7.4-7.6	< 7.2
Pleural/Serum protein ratio	< 0.5	> 0.5
Pleural/Serum LDH ratio	< 0.6	> 0.6

Treatment

- Rx of the cause
- **Chest tube:** Under water seal

Chylothorax

Definition Accumulation of chyle in the pleural space

Etiology

1. **Thoracic duct injury:** Cardiothoracic surgery, chest trauma, neonate (during delivery)
2. **Tumors:** Primary or metastases
3. **Congenital anomalies of lymphatics**
4. **Vascular obstruction (SVC)**
5. **Idiopathic**

Investigations

- CXR
- Thoacocentesis (Milky fluid, may be clear, when?), fluid/serum TAG ratio > 1
- Lymangiogram & lymphoscintigraphy

Treatment

- Low-fat diet, MCT or TPN .
- **Chest tube:** Under water seal
- Octreotide
- Surgical: thoracic duct ligation, pleuroperitoneal shunt...

Pulmonary Tuberculosis

Definition

It is a serious respiratory disease with a mortality rate of 8:100.000 (Egypt)

Etiology

- a. Mycobacterium tuberculosis
- b. Mycobacterium bovis
- c. Mycobacterium africanum

TB Bacilli:

- Aerobic
- Acid-fast bacilli
- Intracellular
- Stained by Ziel-Neelsen stain
- Culture: Lowenstein-Jensen medium

Mode of transmission

- a. Inhalation: Droplet from patient with open TB
- b. Ingestion: Contaminated milk
- c. Inoculation: Skin contact

Pathology (Direct tissue invasion)

1. Tubercle: Caseating granuloma
2. Exudative reaction: Effusion, ascites...

Types & Pathogenesis of TB

1. Primary

- TB infection for the first time
- It occurs in one of 4 sites: Lungs, tonsils, intestine or skin
- Lung is the commonest site to be involved
- Leading to the formation of 1ry complex [Focus, TB lymphangitis, TB lymphadenitis]
- Fate of the 1ry complex:
 - Good immunity: Healing → Fibrosis ± Calcification
 - Poor immunity: Active 1ry disease ± Spread

2. Secondary

- a. Exogenous: Reinfection (Environmental)
- b. Endogenous: Reactivation (TB bacilli that have survived in the 1ry complex)

Primary Pulmonary TB

Etiology

Mode of transmission

1ry complex

1. **Ghon's Focus:** Subpleural, 1-2 cm, usually in the lower part of the upper lobe or upper part of the lower lobe
2. **TB lymphangitis**
3. **TB lymphadenitis** (Hilar LN)

Fate of the 1ry complex

1. Good immunity: Healing → Fibrosis ± calcification (Asymptomatic but reactivation...)
2. Poor immunity: Active 1ry disease ± Spread
 - Direct: Pneumonia, cavitation, pleurisy, pleural effusion
 - Bronchial: Larynx, pharynx, tongue, intestine
 - Blood: Isolated organ or miliary TB
 - LN enlargement: Pressure manifestations
 - Mediastinal structures: Mediastinal syndrome
 - Bronchi: Lung collapse or emphysema

Clinical Picture

A) Symptoms

- Contact with affected family member
- General (TB toxemia): Night fever, night sweat, anorexia, loss of weight
- Chronic cough: is the main symptom with or without expectoration
- Sputum may be mucoid, purulent or bloody sputum
- Chest pain, dyspnea or wheezes may occur
- Any chest symptom with **poor or no response to routine treatment**

B) Signs

- Pneumonia: Signs of consolidation (↓↓ Air entry, bronchial breathing, crepitations)
- Effusion: Stony dullness, ↓↓ Air entry
- Fibrosis: Mediastinal shift to the SAME side (Heart & trachea)
- RD (variable grades 1-4)

Investigations

A) Laboratory

- CBC: Lymphocytosis
- ESR: Markedly ↑↑
- CRP: Positive

B) Imaging

- CXR
- CT chest

C) Bacteriological

- Sample: Sputum, stomach wash, gastric aspirate
- Direct smear: Ziel-Neelsen stain for acid fast bacilli
- Culture: Lowenstein-Jensen medium (requires 3-6 wks to give results)

D) Biopsy

- Sample: LN, pleura, bronchoscopy...
- Pathology: Caseating granuloma

E) Recent methods of diagnosis

- PCR (Polymerase chain reaction)
- ELISA (Enzyme-Linked Immuno-Sorbent Assay)
- BACTEC method: requires 1-3 wks

F) Tuberculin test

- Principle: Delayed hypersensitivity reaction (Type IV) to TB bacilli
- Method: Mantoux test
 - Intra-dermal injection of 0.1 ml containing 5 units of purified protein derivative (PPD) in the skin of the flexor surface of the forearm
 - Area of induration (Not erythema) is measured 48-72 hrs after administration
 - Both longitudinal & transverse diameters are measured (The mean is recorded)
- Interpretation
 - < 5 mm: Negative
 - 5-9 mm: Doubtful (Repeated)
 - > 10 mm: Positive

Positive reaction	False negative reaction
▪ BCG vaccination (usually < 10 mm)	▪ SC administration or Outdated reagent
▪ TB infection	▪ Use of steroids or immunosuppressives
	▪ Cell-mediated immunodeficiency
	▪ Severe malnutrition & cachexia
	▪ Viral vaccines (Measles & mumps)
	▪ Viral infections (Measles & mumps)
	▪ Severe disseminated TB, hypothyroidism

Prevention

A) General measures

- Socioeconomic development & environmental sanitation
- Good housing, proper ventilation & spacing
- Health education
- Proper nutrition
- Food sanitation: Milk sanitation (Pasteurization)
- Regular health appraisal
- Mass radiography centers for early detection of affected individuals
- Diagnosis & Rx of those employed in the care of children in nurseries & schools

B) Specific measures

a. BCG Vaccine [Bacillus-Calmette-Guerin]

- **Nature:** Live attenuated
- **Dose:** 0.1 ml (0.05 ml in neonates)
- **Route:** Intradermal in the Lt deltoid area
- **Timing:** Starting at birth (1st 3 months of life), Booster doses at 5 & 11 yrs
- **Contraindications:** Immunodeficiency, immunosuppressives
- **Precautions:** Tuberculin test is indicated if vaccination is delayed beyond 1st yr
[Only tuberculin-negative individuals are vaccinated]
- **Advantages**
 - Prevention of pulmonary TB ($\approx$ 50% effectiveness)
 - Prevention of TB meningitis & disseminated disease ($\approx$ 50-80% effectiveness)
- **Side effects (Reaction):**
 - Erythema, papule formation $\pm$ ulceration in 3-6 wks
 - Scar formation within 2-6 months
 - Regional lymphadenitis: Anti-TB treatment may be needed
 - Disseminated BCG infections: in immunocompromised individuals

b. Chemoprophylaxis

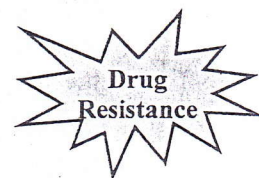
- **Indications:** High-risk close contacts
- **Dose:** Isoniazide (INH) 15 mg/Kg/day for 1 year

Treatment

A) Antituberculous drugs: Combined therapy (2-3 drugs 1st line drugs) for 6-9 months.

Combined therapy is preferred to minimize drug toxicity & development of drug resistance

First Line Drugs	Dose	Route
Isoniazide (INH)	10-15 mg/Kg/day	Oral
Rifampicin (RIF)	10-20 mg/Kg/day	Oral
Pyrazinamide (PZA)	20-40 mg/Kg/day	Oral
Alternative Drugs		
Streptomycin	20-40 mg/Kg/day	IM
Ethambutol	15-20 mg/Kg/day	Oral
Ethionamide	15-20 mg/Kg/day	Oral
Other drugs	Amikacin, Kanamycin, Para-aminosalicylic acid	



B) Steroids may be given, when??

- TB serositis
- TB of the adrenal gland (Replacement therapy)
- TB meningitis
- Allergy to anti-TB drugs
- Miliary TB
- After removal of TB cervical LN to avoid fibrosis
- After removal of TB bronchial LN to avoid stricture

Extrapulmonary TB

A) TB Lymphadenitis

- Cervical LN are the most commonly involved
- Early: Firm, discrete, mobile, non-tender
- Late: Adherent & matted

B) Abdominal TB

a. TB enteritis

- Chronic diarrhea, tenesmus, bleeding
- Stools: Whitish & greasy

b. TB peritonitis

- Ascitic type: Ascites with ↑↑ protein content & ↑↑ cells (mainly lymphocytes)
- Caseous type: Doughy sensation of the abdomen
- Adhesive type: Intestinal loops are glued together by fibrinous adhesions
- Encysted type: Small collections of ascetic fluid between adherent intestinal loops

c. TB mesenteric lymphadenitis (= Tabes mesenterica)

- Enlarged matted mesenteric LN palpable in the Rt iliac fossa with doughy sensation

d. Urogenital TB

- Renal TB results from hematogenous spread
- Affection of renal pelvis, ureters, bladder, prostate, seminal vesicles may follow
- Urine: Sterile pyuria (Culture may reveal TB bacilli)

C) CNS TB

a. TB meningitis

- See before (↑↑ ICP, Irritability, Convulsions...)
- CSF: ↑↑ Protein, ↑↑ cells (mainly lymphocytes), ↓↓ Glucose, ↓↓ Chloride

b. Tuberculoma

- Formation of large caseous lesion
- Space-occupying manifestations

D) Skeletal TB

a. Pott's disease of the spine

- Site: Lower thoracic (*Commonest site*), Lumbar, Cervical
- Nature: Destruction of vertebral bones
- C/P: Angular kyphosis (Not correctable)
- X-rays: Bone rarefaction & destruction

b. TB arthritis

- Site: Hip, Ankle, Knee, Elbow joints
- Nature: Inflammation of the joint
- C/P: Pain, swelling, limitation of movements
- X-rays: Bone rarefaction & destruction

E) Other types

a. TB pericarditis

- Route: Lymphatic spread
- Types (Pathology): Serous & Constrictive
- Diagnosis: Pain, CXR, ECHO

b. TB skin

- Site: Chin, lips, nose, limbs, genitalia (Lupus vulgaris)

c. TB of eye & ear: Conjunctivitis, chronic otitis media, mastoiditis